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# ROENTGENOLOGIC ASPECTS OF CEREBRAL ANGIOGRAPHY IN CHILDREN*

*By* JUAN M. TAVERAS, M.D., *and* CHARLES M. POSER, M.D.†
NEW YORK, NEW YORK

THE value of cerebral angiography as a diagnostic aid in neurology has become well established. The use of the procedure in children is increasing but only a few papers have appeared dealing specifically with cerebral angiography in this age group (Torkildsen and Koppang,[21] Tolosa,[19,20] Picaza,[10] Ramos[14]). In a recent publication,[11] the writers have summarized the clinical aspects of cerebral angiography in children, dealing principally with clinical indications, morbidity and complications. It was shown that cerebral angiography is a relatively harmless procedure, the complications being mild and transient. In our group it was shown to be accompanied by less morbidity than pneumoencephalography performed in a comparable group of children.

It is the purpose of the present communication to review the roentgenologic aspects of cerebral angiography in a series of 138 patients under the age of thirteen, examined at the Neurological Institute of the Presbyterian Hospital.

## MATERIAL

Two hundred and seven cerebral angiographies were performed on 138 children from 1944 through mid-1958. A minimum of 414 separate injections of contrast substance was made. Diodrast was used in 85 patients, urokon in 30 patients, hypaque in 18 and thorotrast in 5. The percutaneous technique was employed in 189 of the 207 angiographies. In one case a catheter was inserted in the brachial artery and in another the material was injected in the antecubital vein. In both cases the filling was less satisfactory than when the conventional technique was used. The age of the patients ranged from five and one-half weeks to twelve years.

Unilateral carotid angiography was performed in 90 patients, bilateral in 40, unilateral carotid and vertebral in 3 and bilateral carotid and vertebral in 8.

## TECHNIQUE

Practically all cases were done by the percutaneous method. Details have been

* From the Departments of Radiology and Neurology, College of Physicians and Surgeons, Columbia University, and the Neurological Institute, Presbyterian Hospital, New York, New York.

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† Present address: University of Kansas School of Medicine, Kansas City, Kansas.

given in a previous publication[11] and we will limit our discussion to the roentgenographic aspects of technique.

Our roentgenographic technique in children is essentially the same as that used in adults. In the past, serial roentgenograms were made at one-half second intervals in frontal and lateral projection for four and one-half seconds, but frequently a second injection with roentgenograms taken during an eight to ten second interval was necessary. For the last three and one-half years we have been making the first six exposures at one-half second intervals. At this point the speed of the serialograph is shifted to 1 per second and four more exposures are made. This allows us to cover seven seconds of circulation time without increasing the exposure to the patient. If there are clinical signs of increased intracranial pressure, which is known to cause lengthening of the circulation time, six exposures at 1 second intervals are taken after the first six made at one-half second intervals. Rarely is there need for repeating the serialograms to cover a longer period of time.

To avoid distortion of shadows it has been our custom to take the lateral roentgenogram with the film against the side being injected. In taking the anteroposterior roentgenograms, it is our aim to direct the roentgen-ray beam in such a way that the superior margin of the orbit and that of the petrous pyramids are superimposed. This projection gives least distortion and maximum visualization of the arteries (Fig. 1 and 5). To accomplish this the roentgen ray tube is angled caudad 12 degrees from the canthomeatal line (external acoustic meatus to outer canthus of the eye) in adults. In children slightly less angulation is necessary; an angle of five degrees is used routinely, except that no angulation is employed in children under five years of age. Roentgenograms in other projections such as the frontal view with more or less angulation of the tube, oblique views and base views are taken only as indicated to study a particular problem.

The lateral serialograms are taken first and then the tube and film changers are moved for the anteroposterior exposures. This maneuver takes approximately five minutes, but ten minutes are usually permitted to elapse between any two injections of the contrast material. It is felt that this reasonable interval between injections should be observed in order to minimize possible complications. The amount of contrast substance (usually 35 per cent diodrast, 30 per cent urokon or 50 per cent hypaque) which was actually injected into children under ten years of age was 6 cc. In those over ten years of age, 8 cc. was used. Since the acquisition of an automatic injector, only 6 cc. is injected in older children and in adults, and 4 cc. in children six years of age or younger. The injection is made as rapidly as possible by hand. With the automatic injector, the injection time is set for approximately 1.25 seconds. Routine compression of the internal carotid artery on the opposite side during the injection of contrast substances has been omitted. Compression is used only when specifically indicated for it is our feeling and that of Decker[3] that routine compression of the opposite side increases the incidence of complications in cerebral angiography.

Vertebral angiography was carried out in 11 cases. Both lateral and frontal serialograms were taken. For the anteroposterior projection, a 25 degree angulation of the tube caudad, relative to the canthomeatal line, is desirable. Occasionally exposures in the base views were made.

## INDICATIONS

These have been reviewed in detail in a previous publication.[11] The main indications in this series of cases were (a) suspected intracranial aneurysm or arteriovenous malformation, (b) suspected supratentorial mass lesion, and (c) vascular occlusion or sinus thrombosis (Table 1).

## ANATOMIC AND PHYSIOLOGIC CHARACTERISTICS

The configuration of the arteries and veins in children is essentially the same as

TABLE I

INDICATIONS AND RESULTS OF CEREBRAL ANGIOGRAPHY IN CHILDREN

(90 patients)

| | No. of Cases | Positive Results | Questionable Results |
|---|---|---|---|
| 1. Suspected Intracranial Aneurysm | 9 | 4 | 1 |
| a. Subarachnoid hemorrhage | 5 | 3 | 1 |
| b. Sudden ophthalmoplegia | 4 | 1 | |
| 2. Suspected Intracranial Lesion (lateralized, supratentorial) | 63 | 30 | 2 |
| a. Suspected neoplasm, abscess or hematoma | 24 | 12 | 1 |
| b. Suspected vascular malformation or neoplasm | 23 | 14 | 1 |
| c. Focal convulsive seizures with suspected fixed lesion | 16 | 4 | |
| 3. Suspected Arterial Occlusion | 12 | 3 | 1 |
| 4. Suspected Sinus Thrombosis | 1 | | |
| 5. Investigative | 5 | | |
| a. Pre- and post-Beck carotid-jugular anastomosis | 4 | | |
| b. Hydranencephaly | 1 | | |
| Total | 90 | 37 | 4 |

in adults. However, there are slight differences in the behavior of the intracranial vessels as well as in the size of the arteries and in the position of the major arterial trunks which might be of interest to review here.

*Contralateral Filling.* Filling of the two anterior cerebral arteries and sometimes of the opposite middle cerebral artery was not uncommon in children even when no compression of the opposite side was used during the injection (Fig. 1). It seemed to be more frequent in younger children. This may be related to the lower blood pressure, since the injection was made with approximately the same force as in adults. The smaller size of the capillary bed may be another factor in younger children if the injection time is relatively prolonged. Still another factor is the use of a greater amount of contrast substance than is actually necessary in the younger group.

*Posterior Cerebral Artery Filling.* This was encountered in a minimum of 40 per cent of the cases. Curtis[1] mentions posterior cerebral artery filling in 28 per cent of cases, Engeset[6] in 30 per cent, and Green and Arana[9] in 34 per cent. These series comprised patients of all ages. In children under ten years of age, some degree of posterior cerebral filling occurred in about 50 per cent of angiographies.

*Circulation Time.* Arterial circulation

FIG. 1. Normal anteroposterior angiogram of a twenty-four month old child. There is bilateral filling although no compression of the opposite side was used during injection. Upper margins of orbits and petrous pyramids are superimposed (dotted lines). Note symmetric distribution of the middle cerebral branches over the island of Reil.

TABLE II

ARTERIAL CIRCULATION TIME

(Estimated only in normal angiograms)

| Time (seconds) | No. of Cases | Per Cent of Cases |
|---|---|---|
| 1.0 to 1.25 | 17 | 32.0 |
| 1.5 to 1.75 | 23 | 43.4 |
| 2.0 | 9 | 17.0 |
| 2.5 | 4 | 7.5 |
| Total | 53 | 100.0 |

Average arterial phase—1.5 seconds.

time was tabulated in 53 patients who were considered to have normal angiograms and the results are as shown in Table II. In 10 of these cases the injection was made with the automatic injector. The arterial circulation time was considered as the time necessary for the contrast substance to go from the carotid siphon to the point where only terminal minor arterial branches remained filled. It included both the "arterial filling" and the "arterial emptying" phases as described by Greitz.[8] The length of the arterial phase cannot be shorter than the injection time and it is possible that the speed of injection may have influenced the more prolonged arterial phase (2.5 seconds) noted in 4 of the cases. This factor can only be eliminated when a relatively small amount of contrast material (2 or 3 cc.) is injected by means of an automatic injection device with the needle correctly placed in the arterial lumen. There were 5 patients in whom both sides were examined at the same sitting. Some discrepancy in the arterial circulation time was found when one side was compared to the other. In 2 patients who had a relatively long arterial phase (2.5 seconds), the time was equal on both sides. This tends to indicate a physiologic condition rather than an artefact due to a slow injection.

The average arterial circulation time was 1.5 seconds if all cases are included. When only the 10 normal cases in which the automatic injector was used are considered, the average arterial circulation time was 1.36 seconds. The difference is probably not significant.

The so-called "capillary phase" is usually not as well defined in children as in adults. The venous phase (measured from the time of the appearance of veins to complete venous emptying) could not be estimated accurately because in most cases a complete venous phase was not recorded. In those cases where it was recorded, it varied from 3.0 to 7.25 seconds. In one case it was 1.5 seconds. These figures are not different from those in the adult. It is impossible to define accurately the end of the venous phase because the contrast substance fades gradually and at different times in different veins.

The *circulation time* as determined by the method suggested by Greitz[8] (time elapsed from maximum concentration in the carotid siphon to maximum concentration of the opaque substance in the parietal veins) was found to vary from 1.5 to 5.0 seconds. The average in 27 arteriographies was 3.3 seconds. The average circulation time as published by Greitz in 32 adults was 4.13 seconds. It is probable that circulation time estimated in this manner is slightly shorter in children than in adults. Since the longest circulation time was 5.0 seconds, any child with a circulation time of 5.5 seconds or over should be considered as abnormal. The most frequent cause of an abnormally prolonged circulation time was increased intracranial pressure; another cause was arterial thrombosis.

The configuration of the superficial cerebral veins is essentially the same in children as in adults. The forward curve of the superior superficial cerebral veins as they turn rostrad to enter the superior longitudinal sinus was less marked in children. This is probably related to brain growth.

*Caliber of the Arteries.* The caliber of the branches of the internal carotid artery on cerebral angiograms, as measured at several pre-determined points, in children was slightly larger in the average than in adults with normal angiograms. Only adults be-

tween twenty and forty-five years of age were chosen for comparison in order to eliminate arteriosclerosis as a possible source of error. To measure the caliber of the vessels, a magnification technique was used; the films were projected over a smooth screen. This method suffers from certain inaccuracies chiefly due to the penumbra effect—a common source of error in all measurements of small roentgenographic shadows—but the error was considered to be essentially equal in all cases. It is difficult to obtain comparable measurements of vessels in different patients. We decided to take five measurements: one of the carotid siphon, two of the anterior cerebral arteries and two of the middle cerebral arteries (in the middle cerebral artery the largest branch was used) at specified distances from the carotid bifurcation (Fig. 2). All measurements were taken twice without any knowledge as to whether a child or an adult was being measured. The figures obtained from the various measurements in 30 children and 37 adults with normal angiograms were submitted to statistical analysis and it was found that the differences encountered were significant. The arteries were found to be larger in children, particularly the proximal portion of the anterior and middle cerebral arteries corresponding to "B" and "D" in Figure 2. The measurements were larger even when children of all ages were included, but if children under five years of age were excluded the discrepancy was greater. We feel that the difference in the size of the vessels is most probably due to some degree of diffuse vascular spasm present in adults during angiography. Since the same drug (chiefly diodrast) was used in both groups of cases, the difference cannot be ascribed to drugs. Practically all of the angiographies in children were performed under general anesthesia. Whether general anesthesia decreases cerebral vascular spasm or whether the larger caliber is related to a less complete development of the autonomic control or to greater elasticity of the vascular walls in children cannot be

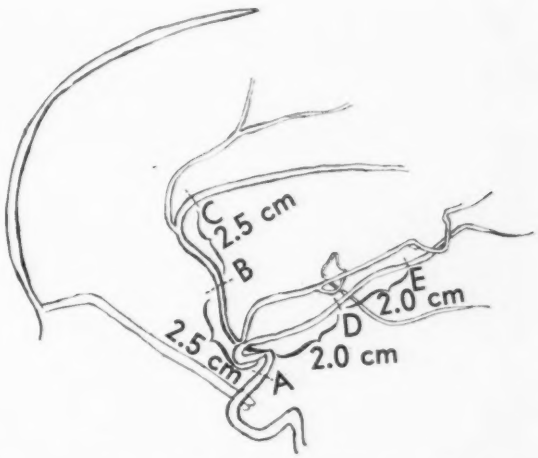



Fig. 2. Diagram illustrating points at which measurements were taken in the carotid siphon, in the anterior and in the middle cerebral arteries.

stated at present. It might also be related to a relatively greater demand for oxygen in children. We have had occasion to see a number of adult patients in whom the vascular spasm, associated with the presence of an aneurysm which recently bled, was not modified by general anesthesia. Anesthesia was necessary when the patient became uncooperative after one or two injections of the contrast material. Similarly, in a few patients who presented evidence of marked vascular spasm a second injection was made with thorotrast without any appreciable change in the spasm, thus eliminating diodrast as a cause of the spasm in these patients. Children, as well as adults, with bleeding aneurysms show local and diffuse vascular spasm which is not modified by general anesthesia or by the type of contrast substance used (Fig. 16).

*Relationship between Middle Cerebral Artery and the "Clinoparietal Line."* It has been our impression that the middle cerebral artery, in the lateral projection, frequently appears to be elevated, particularly in young children, in an otherwise perfectly normal arteriogram. This is especially important in cases where slight rotation of the head increases this impression of elevation sufficiently to suggest a temporal lobe mass.

The middle cerebral artery bears a

FIG. 3. Same case as in Figure 1. Normal lateral angiogram showing relationship of lowest main branch of the middle cerebral artery to clinoparietal line (line traced from the anterior clinoid process to a point 2 cm. above the lambda). The lambda is indicated by an arrow.

fairly constant relationship to a line drawn from the anterior clinoid process to a point 2 cm. above the lambda. The middle cerebral artery of normal children is always above this line (Fig. 3). In adults the lambda is often not visible and, if so, a point 9 cm. above the internal occipital protuberance, measured on a straight line is used.

The distance between this line and the middle cerebral branches is measured 2 cm. posterior to the point where the line intersects the internal carotid artery (Fig. 3). The measure is more accurate in children than it is in adults for, as pointed out by Torkildsen and Koppang,[21] the middle cerebral vessels are less tortuous in children.

This relationship has been studied in 73 normal lateral arteriograms. Figure 4*A* shows that the average distance between the middle cerebral artery and the clinoparietal line becomes shorter with advancing age, gradually approaching the adult normal. The average "normal" distance in children lies between 5 and 16 mm. Figure 4*B* shows the measurements of the distance of the clinoparietal line and the middle cerebral artery in 65 normal adults. No definite trend can be noted. The longest distance encountered was 9 mm. *above* the line and the shortest 4 mm. *below* it. Out of 65 adult patients, ranging in age from twenty-two to sixty-two, in only 5 was the middle cerebral artery below the line, and in 6 it was 7 to 9 mm. above it. In 54 (83 per cent) it was from 0 to 6 mm. above the line.

A comparison of the middle cerebral artery—clinoparietal line distance meas-

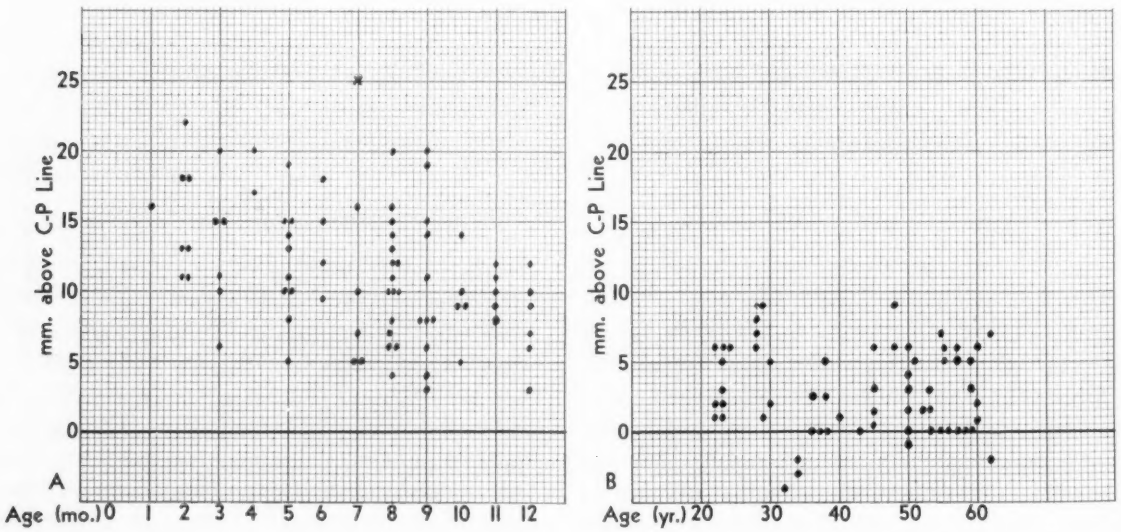



FIG. 4. Distance between lowest main branch of the middle cerebral artery and the clinoparietal line. (*A*) In children; (*B*) in adults.

ured on both sides in the same patient was made in 20 children. A slight difference in the height of the vessels when the right was compared with the left was found in most patients. The difference was usually small and ranged between 1 and 5 mm. In only 4 cases was it above 5 mm. The largest difference in a normal person (confirmed by air study) was found to be 11 mm. This difference was thought not to be due merely to projection, but to actual difference in the height of the vessels. The importance of this difference in the position of the middle cerebral artery when the right side is compared with the left is obvious, and realization that this constitutes a normal variation will prevent an erroneous diagnosis of temporal lobe mass lesions.

The lowering in the position of the middle cerebral artery with advancing age is evidently related to the growth of the brain. The sylvian fissure becomes lower with advancing age probably because of the relatively greater growth of the frontal lobes. Another finding related to brain growth is that the high position of the anterior cerebral artery in children often gives the impression of a suprasellar mass (Fig. 5).

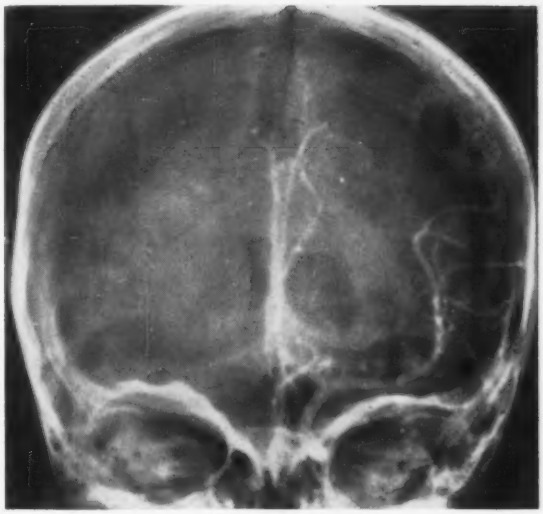


Fig. 5. Normal anteroposterior angiogram of a five year old child showing apparent elevation of the anterior cerebral artery. This is occasionally seen in normal adults but is more common in children.

## ROENTGEN FINDINGS

Cerebral angiography when properly indicated is of value in the diagnosis of intracranial disease in children.

Of 132 patients in our series, angiography was helpful in obtaining information which tended to elucidate the clinical problem in 58. The over-all positive result in this series was 43 per cent (we have eliminated 6 cases in which angiography was performed for purely investigative reasons). The lesions most frequently diagnosed by angiography in adults are supratentorial neoplasm and saccular aneurysm. In children the proportions are different; venous and arteriovenous angiomas and subdural hematomas were encountered as frequently as neoplasms. There were only 2 saccular aneurysms out of the 138 children, a rather small number for a lesion which is usually assumed to be congenital (Table III). Perhaps the defect in the artery exists without the aneurysmal sac. It is difficult to be certain of this. The relatively low in-

Table III

ROENTGENOLOGIC FINDINGS IN CEREBRAL ANGIOGRAPHY IN 132 CHILDREN

| Diagnosis | No. of Cases |
|---|---|
| Vascular malformation | 15 |
| Carotid-cavernous fistula (post-traumatic) | 1 |
| Cerebral atrophy | 10 |
| Subdural hematoma | 6 |
| Neoplasm | 10 |
| Ventricular dilatation | 6 |
| Mass lesion (cyst or hematoma) | 3 |
| False aneurysm | 1 |
| Arterial thrombosis | 4 |
| Cerebral abscess | 2 |
| Congenital arterial aneurysm | 2 |
| Angioma and arterial thrombosis | 1 |
| Angioma and sinus thrombosis | 1 |
| Angioma and diffuse cerebral atrophy | 1 |
| Ventricular dilatation and tumor | 1 |
| No definite diagnosis—abnormal angiogram | 4 |
| Hydranencephaly | 1 |
| Carotid-jugular fistula, surgical | 4 |
| Normal | 59 |
| Total | 132 |

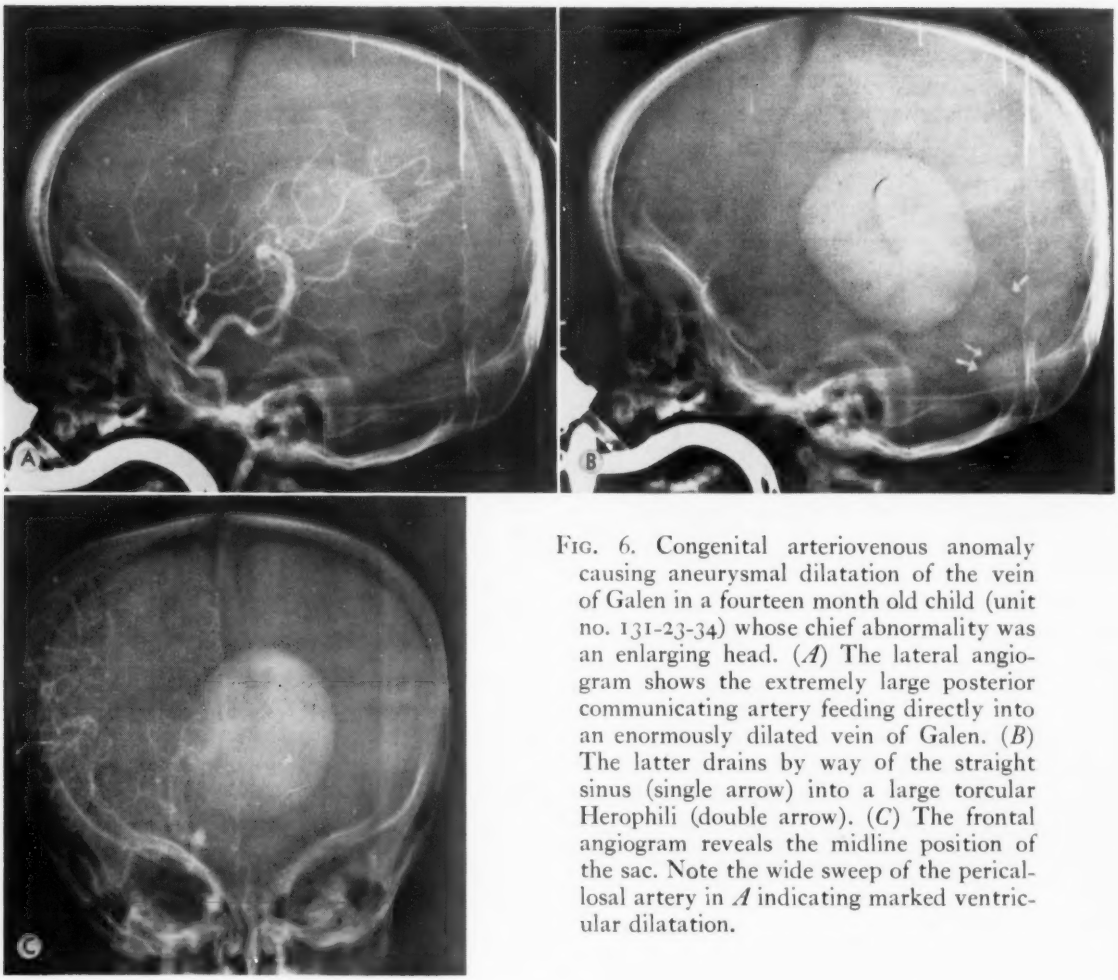


FIG. 6. Congenital arteriovenous anomaly causing aneurysmal dilatation of the vein of Galen in a fourteen month old child (unit no. 131-23-34) whose chief abnormality was an enlarging head. (*A*) The lateral angiogram shows the extremely large posterior communicating artery feeding directly into an enormously dilated vein of Galen. (*B*) The latter drains by way of the straight sinus (single arrow) into a large torcular Herophili (double arrow). (*C*) The frontal angiogram reveals the midline position of the sac. Note the wide sweep of the pericallosal artery in *A* indicating marked ventricular dilatation.

cidence may be due to the fact that bleeding occurs less frequently in children.

In this series there were 15 children who had a clinical picture compatible with the diagnosis of Sturge-Weber syndrome. Several different types of vascular abnormalities were encountered: venous "angiomas" (telangiectasis), capillary "angiomas," arterial thromboses, abnormal veins, thrombosis of the superior longitudinal sinus, and superficial angiomas in the scalp supplied by the internal carotid system. A detailed description and illustration of these abnormalities is not justified here and has been reported separately.[12] Serialograms are, of course, indispensable in the study of this syndrome or whenever an arteriovenous malformation is suspected.

In some patients in whom an arteriovenous malformation was demonstrated on the angiogram, a pneumoencephalogram showed local cerebral atrophy (porencephalic dilatation of the ventricle). For this reason, if the air study is done first, angiography should be performed when local or unilateral cerebral atrophy is encountered. At other times the malformation presented as a mass lesion and produced displacement of the ventricles or blockage of the flow of the cerebrospinal fluid (Fig. 6).

*Cerebral Atrophy*. Diffuse cerebral atrophy or hypoplasia may be suspected but not diagnosed angiographically, and cerebral hemiatrophy may be diagnosed by angiography, although it is well known that pneumoencephalography is the diagnostic procedure of choice in these condi-

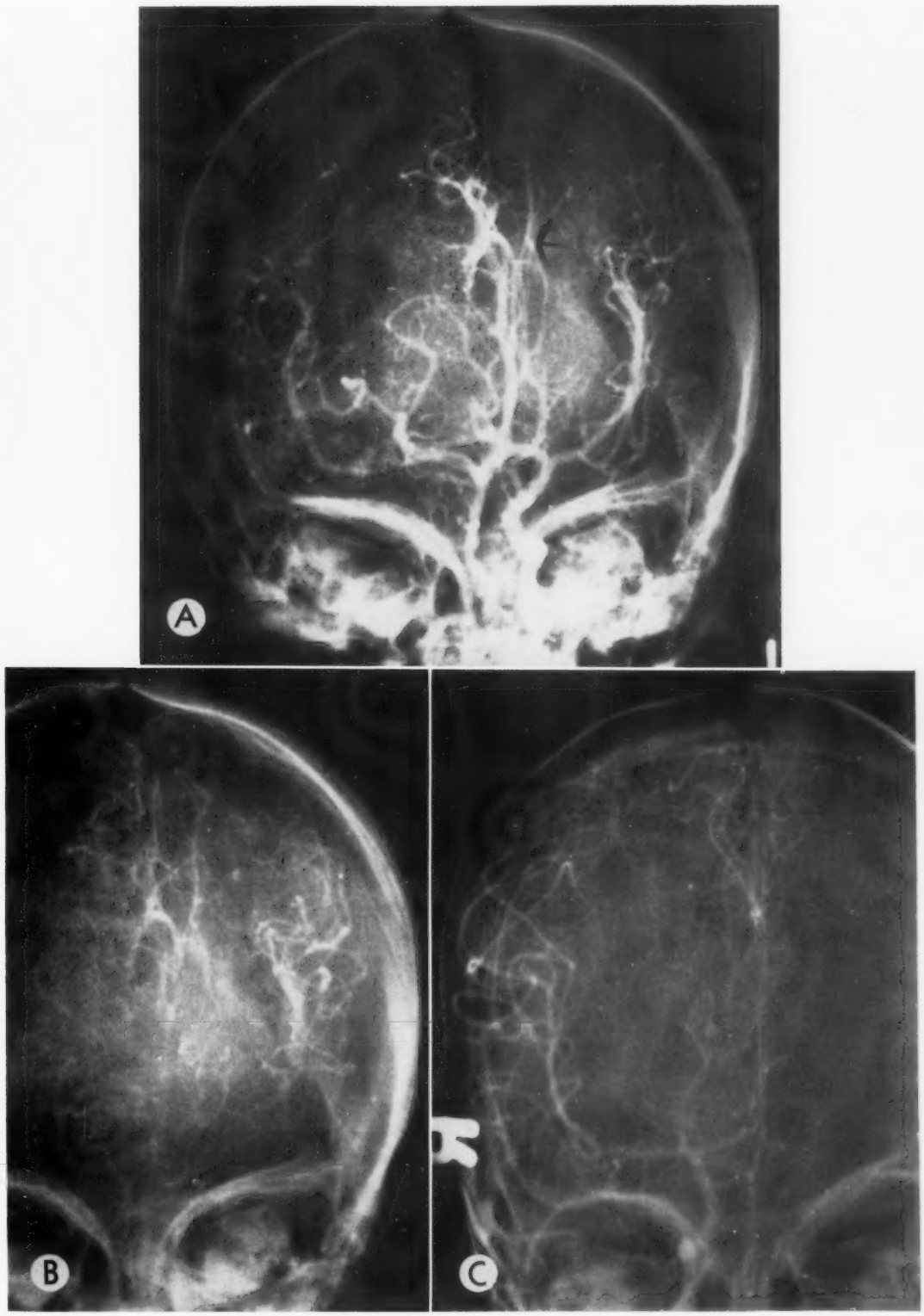



FIG. 7. Cerebral atrophy or hypoplasia demonstrated angiographically in a twenty-seven month old child (unit no. 14-23-24) with encephalo-trigeminal angiomatosis. (*A* and *B*) Note the great distance between the surface vessels and the inner table of the skull, bilaterally. (*C*) Cerebral atrophy in a two year old child (unit no. 13-52-85). There is a uniform separation of 4 mm. between the branches of the middle cerebral artery and the inner table of the skull. (Same case as in Figure 18.)

tions. The angiographic findings in diffuse cerebral atrophy, where there is an increased amount of fluid in the subarachnoid space, consist of a uniform separation of the cortical vessels—arteries as well as veins—from the inner table of the skull. Normally, there is a separation of only 1 to 2 mm. between the vessels and the inner table. In the presence of cortical atrophy, the distance is increased to 4 or 5 mm. or more. The uniformity throughout the hemisphere and the rather slight separation should make one reluctant to diagnose a subdural hematoma (Fig. 7).

A diagnosis of cerebral hemiatrophy can be made accurately by angiography. Normally, there is a distance of over 2 cm. between the inner table of the skull and the innermost limit of the middle cerebral vessels over the insula (island of Reil) and just before they leave the insula to emerge through the sylvian fissure and over the surface of the hemisphere. Two measurements were taken as indicated in Figure 8. In a group of 30 normal angiograms the minimum distance for "A" was found to be 21 mm., and the maximum distance 30 mm. The measurement of "B" (in Fig. 8) gave a minimum of 28 mm. and a maximum of 40 mm. The distance between the middle cerebral artery and the inner table (Point A in Fig. 8) in well established cases of cerebral hemiatrophy was always less than the minimum normal and ranged between 14 and 19 mm. The latter point would be of help in differentiating between hemiatrophy and a mass in the opposite side (Fig. 9). In one case of relatively recent onset, the measurement was at the lowermost limit of normal (Fig. 11). In the presence of cerebral hemiatrophy, there is a deviation of the anterior cerebral artery to the atrophic side. The anterior cerebral vessels ascend straight up and do not show a second deviation at the lower edge of the falx cerebri because the falx is also situated off center towards the atrophic side. This feature differentiates it from midline shift due to a mass lesion (Fig. 9). While many cases of cerebral hemiatrophy are the result of severe trauma at birth or early in childhood with damage to the vessels,[5] this is not an indispensable factor and may actually be of secondary importance or absent altogether from the history. In our series there were 8 patients with cerebral hemiatrophy. Thrombosis of the middle cerebral artery was encountered in only 1 of the 8 cases—a child who also had the Sturge-Weber syndrome—but several children revealed a diminution in the caliber and in the number of the middle cerebral branches in relation to that of the anterior cerebral artery. The smaller caliber suggests that vascular injury or actual thrombosis took place at an earlier age followed by recanalization (Fig. 10 and 11). We have not

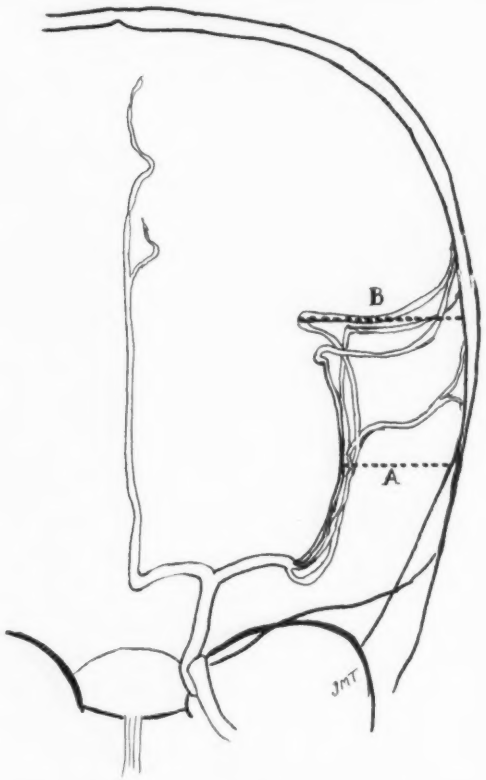



FIG. 8. Diagram illustrating the points at which measurements from the middle cerebral artery branches to the inner table of the skull were taken. The distance corresponds to the thickness of brain tissue, the island of Reil to the surface of the brain. A decrease in this distance may indicate either atrophy of the brain or lateral displacement due to ventricular dilatation or a deeply situated tumor.

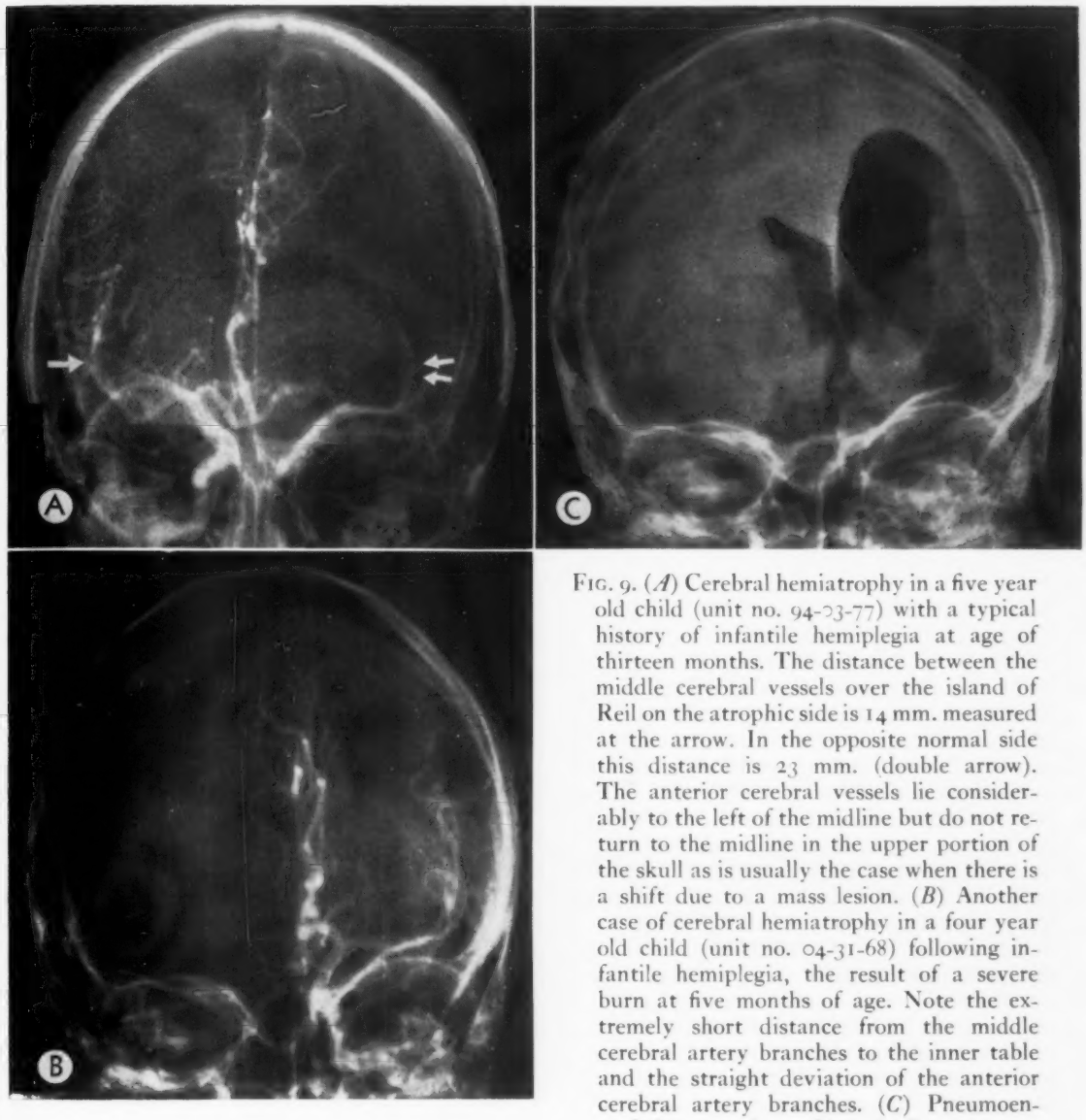



Fig. 9. (*A*) Cerebral hemiatrophy in a five year old child (unit no. 94-03-77) with a typical history of infantile hemiplegia at age of thirteen months. The distance between the middle cerebral vessels over the island of Reil on the atrophic side is 14 mm. measured at the arrow. In the opposite normal side this distance is 23 mm. (double arrow). The anterior cerebral vessels lie considerably to the left of the midline but do not return to the midline in the upper portion of the skull as is usually the case when there is a shift due to a mass lesion. (*B*) Another case of cerebral hemiatrophy in a four year old child (unit no. 04-31-68) following infantile hemiplegia, the result of a severe burn at five months of age. Note the extremely short distance from the middle cerebral artery branches to the inner table and the straight deviation of the anterior cerebral artery branches. (*C*) Pneumoencephalogram of case shown in *B*.

had the opportunity to repeat angiography after a long interval following the visualization of middle cerebral artery thrombosis in children in order to demonstrate the occurrence of recanalization. However, we have had occasion to see narrowing and a decrease in the number of middle cerebral branches in a child several months after a typical history of an infantile hemiplegia while hemiatrophy was developing (Fig. 11).

*Arterial Thrombosis.* There were 3 patients with infantile hemiplegia due to thrombosis of the middle cerebral artery demonstrated angiographically (Fig. 12). Two other cases showed paucity of vessels in the middle cerebral territory believed to be due to recanalization (Fig. 10 and 11). Ten other cases with the same clinical picture were examined but the angiogram was considered normal. In these cases there may have been thrombosis of smaller branches which was not visualized on the angiogram.

*Subdural Hematoma.* There were 5 cases of subdural hematoma. Of these, 4 were of the so-called juvenile relapsing type.[2] They

FIG. 10. Old middle cerebral artery thrombosis showing paucity of branches in the middle cerebral distribution. The ramifications of the middle cerebral artery are considerably smaller than the anterior cerebral artery and its branches. The child was an eight year old girl (unit no. 17-92-29) with a history of focal right sided seizures for about a year. There were only slight neurologic signs involving the right extremity, indicating an old hemiparesis.

showed evidence of enlargement of the middle fossa on the side of the hematoma and also elevation of the sphenoid ridge (lesser wing of sphenoid) as seen through the orbits in Caldwell's projection (Fig. 13). An angiogram showed findings typical of subdural hematoma, the largest portion of which was in the temporal fossa.

*Vascular Malformations.* There were 15 cases of vascular malformations, some in the posterior fossa and some in the supratentorial region. Of the latter group, in 3 cases the blood was shunted directly into the deep veins and straight sinus. In 2 of these, there was an aneurysm of the vein of Galen, which was actually an arteriovenous communication with a saccular dilatation of the major draining vein (Fig. 6). This type of anomaly can easily be confused with a third ventricle neoplasm on air studies. Indeed, this was the case in a fourth instance where a round mass was encountered on the superior and posterior portion of the third ventricle, producing moderate internal hydrocephalus in a six month old child. Autopsy two years later

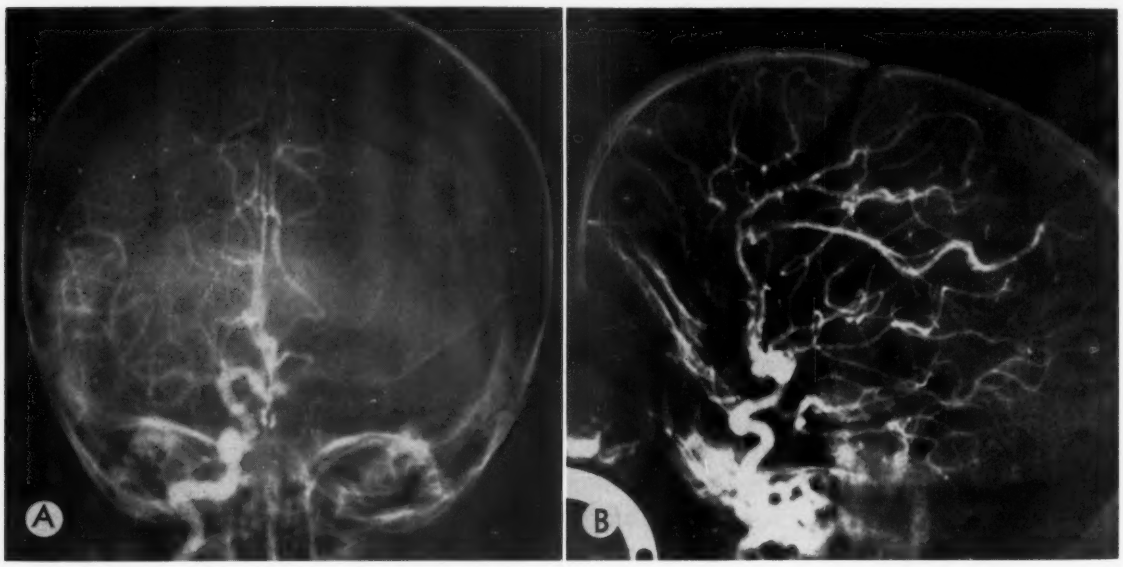



FIG. 11. Cerebral hemiatrophy in a thirteen month old child (unit no. 13-17-37). Six months prior to arteriography the infant had developed repeated left sided convulsive seizures followed by left hemiparesis which persisted. (*A*) Note straight deviation of anterior cerebral branches. The distance from the middle cerebral artery group to the inner table is 21 mm., which is at the lower limit of normal, but might become smaller as time goes on. (*B*) There is relative paucity of arterial branches in the middle cerebral artery distribution in comparison to the anterior cerebral group, indicating an old thrombosis of the middle cerebral artery probably followed by recanalization.

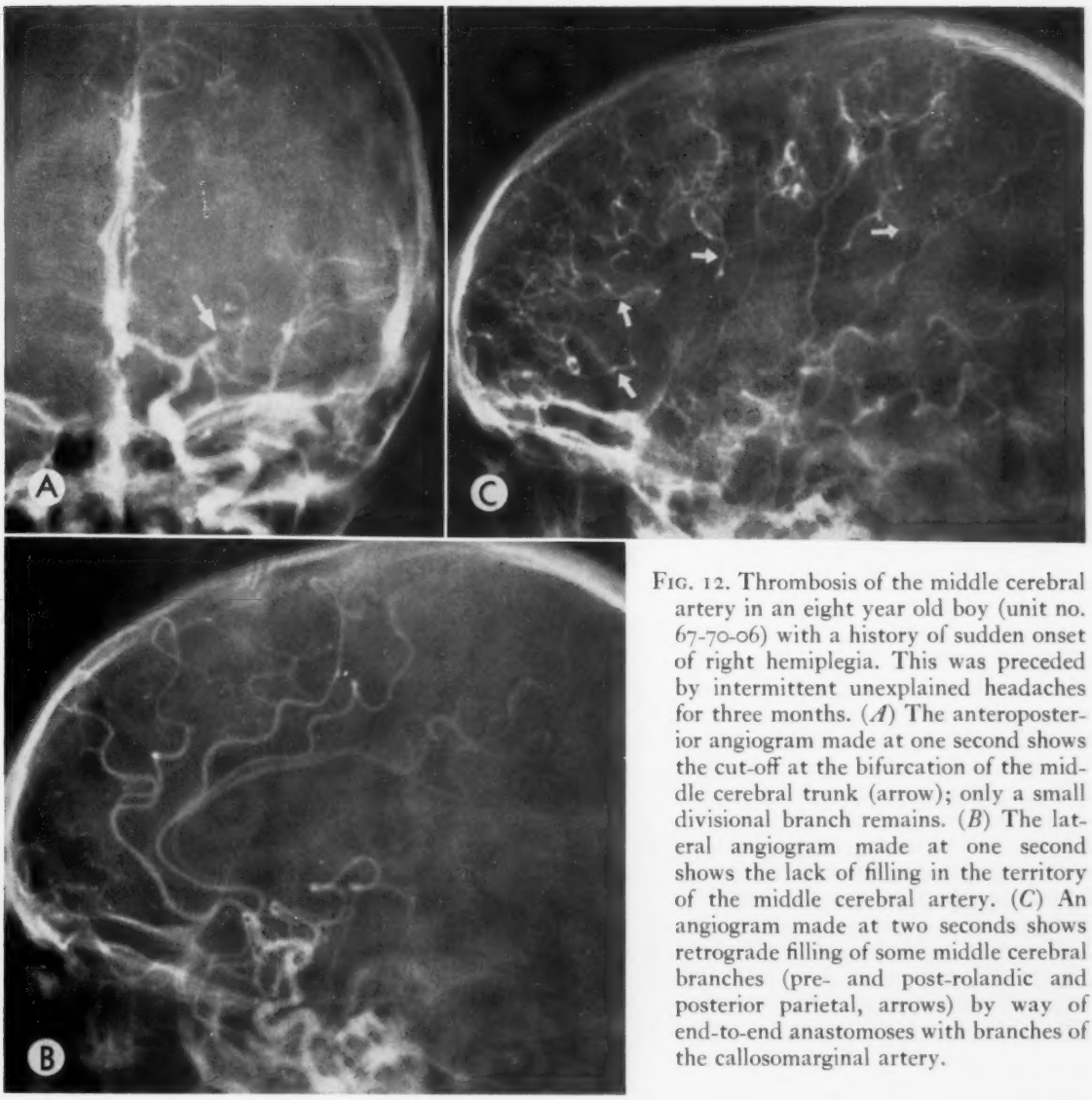



Fig. 12. Thrombosis of the middle cerebral artery in an eight year old boy (unit no. 67-70-06) with a history of sudden onset of right hemiplegia. This was preceded by intermittent unexplained headaches for three months. (*A*) The anteroposterior angiogram made at one second shows the cut-off at the bifurcation of the middle cerebral trunk (arrow); only a small divisional branch remains. (*B*) The lateral angiogram made at one second shows the lack of filling in the territory of the middle cerebral artery. (*C*) An angiogram made at two seconds shows retrograde filling of some middle cerebral branches (pre- and post-rolandic and posterior parietal, arrows) by way of end-to-end anastomoses with branches of the callosomarginal artery.

disclosed the true nature of the mass.

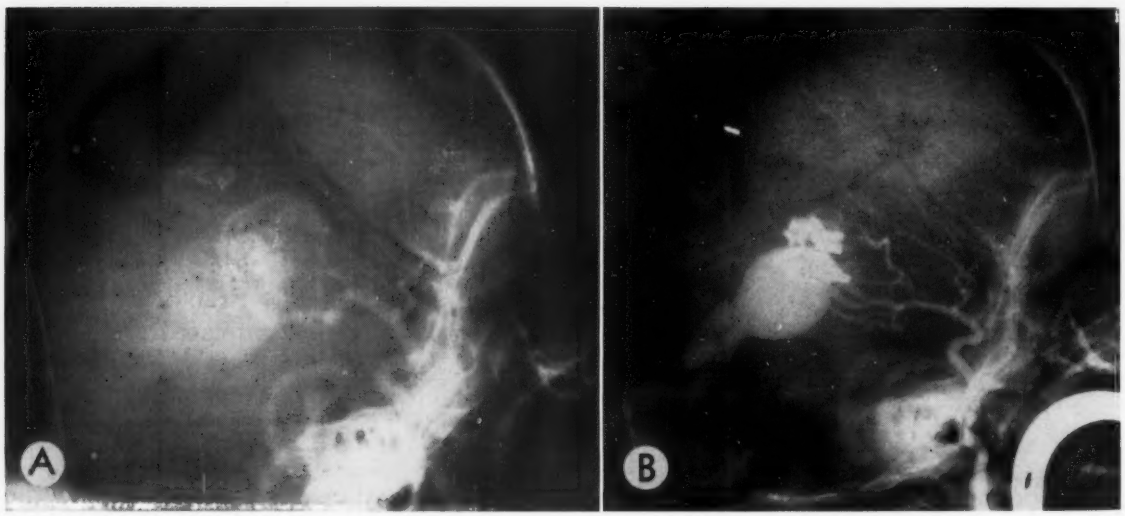
In one of the patients there was enlargement of the heart which was thought to be related to the arteriovenous communication. Repeat chest roentgenograms taken five weeks following an attempt surgically to clip the feeding vessels showed that the heart was smaller (Fig. 14). Repeat angiography at that time revealed a considerable decrease in the size of the venous sac of the aneurysm, indicating a corresponding decrease in the blood flow through it. The decrease in the size of the heart is of interest because, as reported by Schlesinger and Hazen,[15] there is usually no evidence of cardiac enlargement in adults with large arteriovenous malformations in the brain. This might be related to hydrodynamic principles, the head being higher than the heart. In infants who are most often in a horizontal position during angiography, however, enlargement of the heart may occur. Or perhaps this is related to the relatively larger percentage of the blood volume which passes through a large arteriovenous fistula in an infant. Epstein *et al.*[7] have demonstrated that there is a correlation between the size of the shunt and the

FIG. 13. Juvenile "relapsing" subdural hematoma in a twelve year old boy (unit no. 11-65-25). (*A*) The posteroanterior roentgenogram made in Caldwell's projection shows typical elevation of the sphenoid ridge on the left side. (*B*) An angiogram reveals the displacement of middle cerebral branches away from the inner table, more pronounced in the temporal region.

increase in cardiac output. Silverman and co-workers[17] report two instances of heart failure followed by death in very young infants. Our case is of sufficient interest to be reported briefly in this paper.

M.F., a seven month old infant (unit no. 1375311) was brought to the hospital because of an enlarging head and prominence of neck veins. The infant could not hold her head up.

FIG. 14. Congenital arteriovenous malformation draining by way of the vein of Galen and causing marked dilatation of it. (*A*) There is extremely rapid filling of the aneurysmal sac; the serialograms indicated a circulation time of 0.75 seconds. (*B*) Following surgical intervention there was a remarkable decrease in blood flow (8 to 10 times less than previously) as evidenced by a decrease in the size of the internal carotid artery to half its previous size, and by a lengthening of the circulation time to 1.75 seconds (see case report). Note also improvement in cerebral circulation.



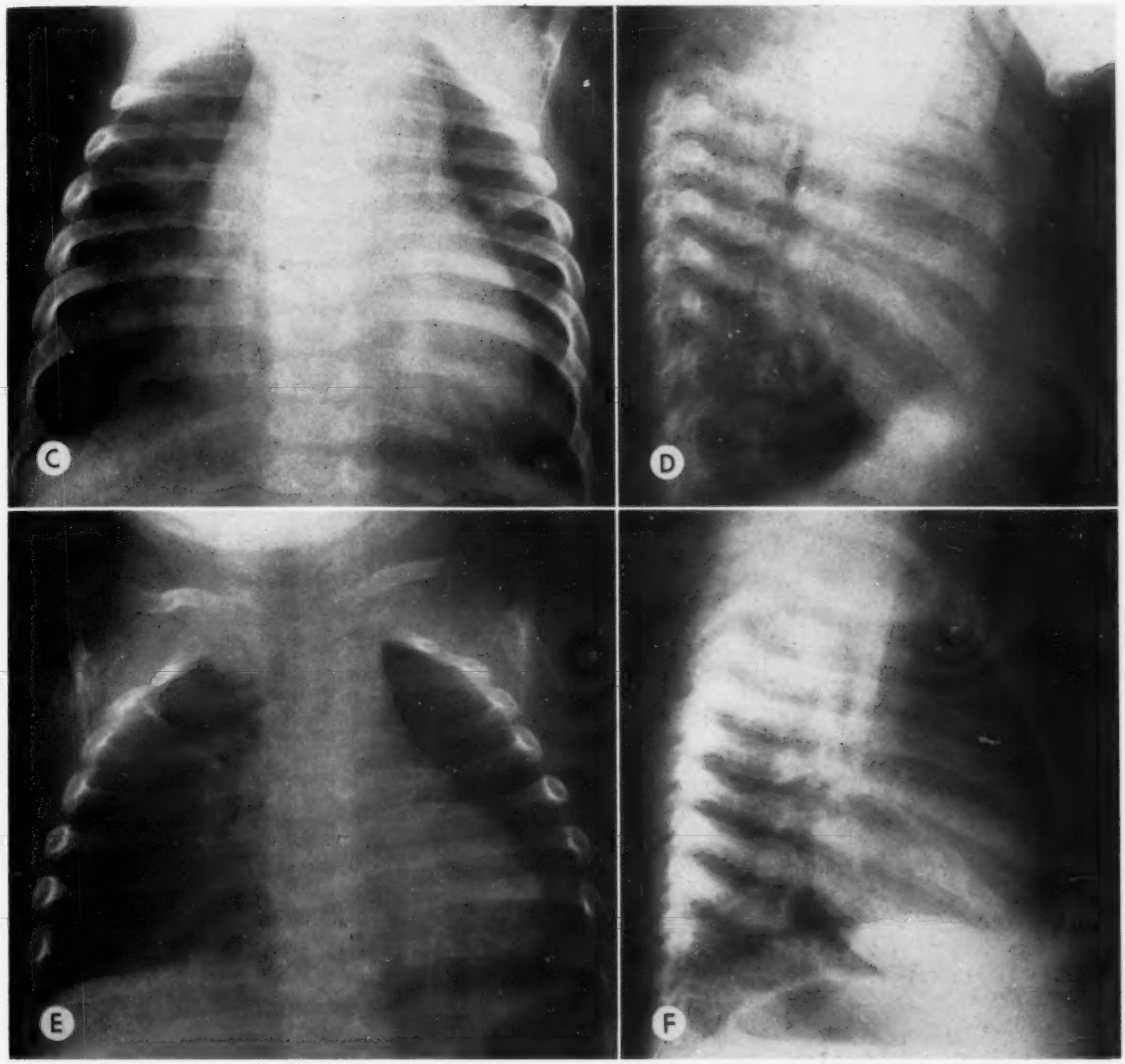



Fig. 14. (*C–F*) Roentgenograms of the chest made before and after operation demonstrating the decrease in the size of the cardiac shadow. The cardiac thoracic ratio in *C* is 0.65 and in *E* 0.57. Note that in *D* the sternum bulges forward and the cardiac shadow overlaps the spine. In *F* the appearance is more nearly normal. The wide upper mediastinum may be produced by a large thymus, but it is thought that the heart shadow has become significantly smaller, particularly as noted in the lateral view. Note also that the preoperative chest roentgenogram (*C*) is in a better inspiratory phase than *E*, which would tend to minimize the actual difference.

All extremities were somewhat spastic; a bruit was heard over the right eye. There was a Grade II systolic murmur over the precordium.

An arteriogram showed a large arteriovenous fistula draining by way of the vein of Galen (Fig. 14*A*). A chest roentgenogram at this time demonstrated an enlarged heart (Fig. 14, *C* and *D*).

An attempt was made surgically to ligate the feeding arteries.

A repeat angiogram showed considerable decrease in blood flow through the fistula (Fig. 14*B*), resulting in pronounced diminution in the size of the sac to less than one-third its previous volume. The caliber of the internal carotid artery decreased fully 50 per cent, which would result in a 75 per cent decrease in flow assuming the same speed of circulation. However, the serialograms indicated an increase in the circulation time (measured from maximum concentration in the carotid siphon to maximum concentration in the surface veins) from 0.75

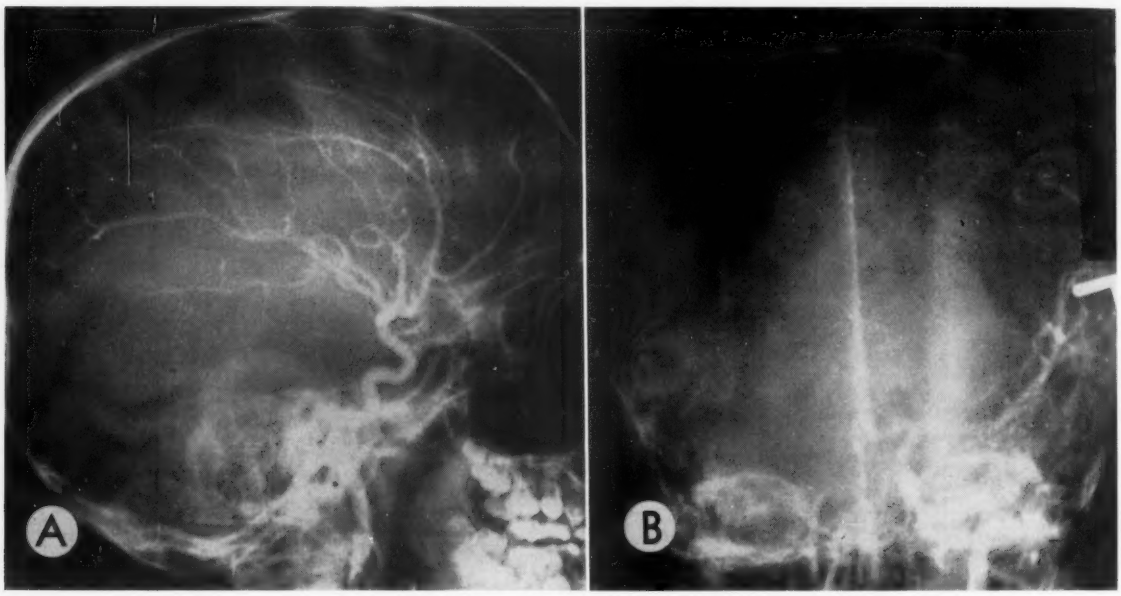



FIG. 15. Ventricular dilatation in a twelve year old child (unit no. 25-69-22) due to third ventricle tumor. (*A*) Note wide sweep of pericallosal artery, elevation and stretching of middle cerebral group. (*B*) There is outward displacement of middle cerebral vessels resulting in widening of space between midline and middle cerebral arteries.

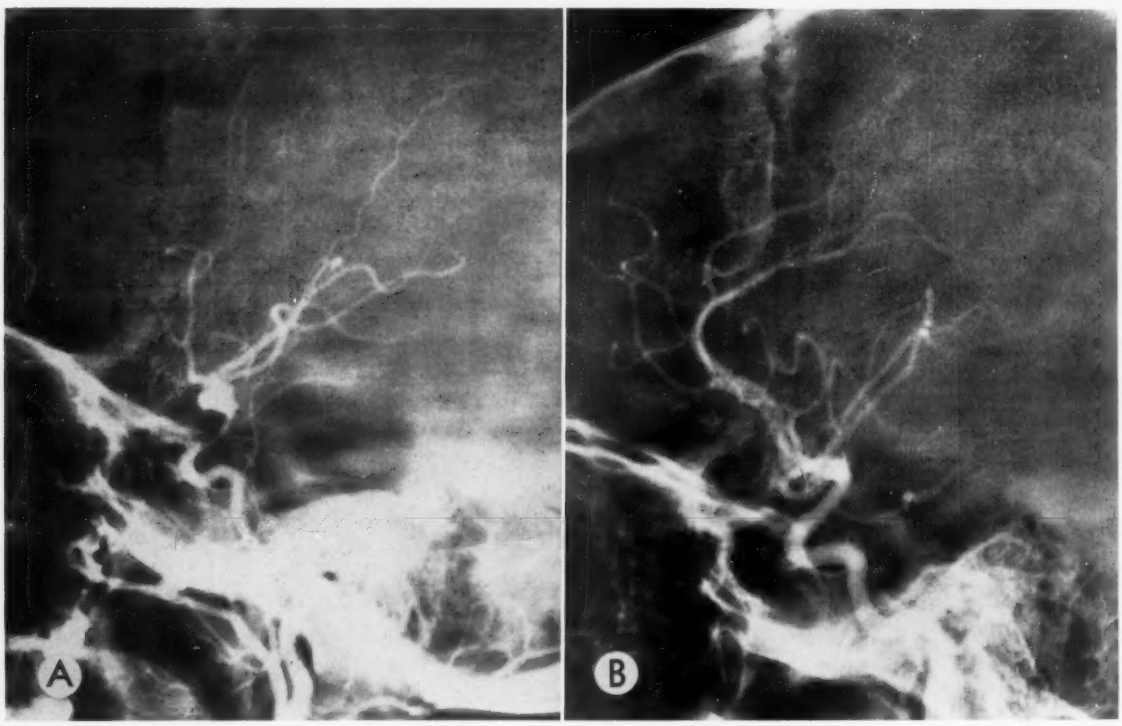



FIG. 16. (*A*) Marked narrowing of the supraclinoid portion of the carotid siphon believed to be due to spasm secondary to a bleeding aneurysm at the bifurcation of the internal carotid artery. The patient was an eleven year old girl (unit no. 22-94-17) with a typical history of subarachnoid hemorrhage a week before. (*B*) An angiogram of the opposite side revealed no evidence of spasm.

seconds before operation to 1.75 seconds after operation. The doubling of the circulation time multiplied the reduction in blood flow. The total decrease in flow probably ranged between 8 and 10 times less flow through the fistula after operation. There was also improvement in the circulation of the hemisphere as evidenced by better filling of the middle cerebral artery and its branches.

A repeat chest examination five weeks after surgical intervention revealed a marked decrease in the size of the heart (Fig. 14, *E* and *F*).

*Hydrocephalus.* In congenital internal hydrocephalus there is usually considerable straightening and stretching of the branches of the internal carotid artery. The sweep of the pericallosal artery is wider than usual. In the frontal projection the carotid siphon and its bifurcation into middle and anterior cerebral arteries have a "Y" configuration instead of the usual "T".[19,20] The vessels are narrower than usual due to stretching and increased intracranial pressure. In acquired hydrocephalus the appearance is somewhat similar but the stretching is not generally as marked (Fig. 15).

*Attenuation of Arterial Lumen.* As mentioned above, arterial spasm associated with a bleeding intracranial aneursym is a frequent occurrence in adults. It also occurs in children (Fig. 16). The spasm manifests itself as a local or diffuse narrowing of the vessels, most often in the neighborhood of the aneurysm and on the same side. Less commonly, it involves a wider group of arteries on the side of the bleeding aneurysm, and still less frequently local arterial narrowing is seen on the opposite side.

The presence of blood in the subarachnoid space might potentiate the appearance of spasm of the cerebral arteries as demonstrated angiographically, for in the series of cases reported by Taveras, Fletcher and Pool[18] there was a correlation between the incidence of spasm and the presence of blood in the cerebrospinal fluid on lumbar

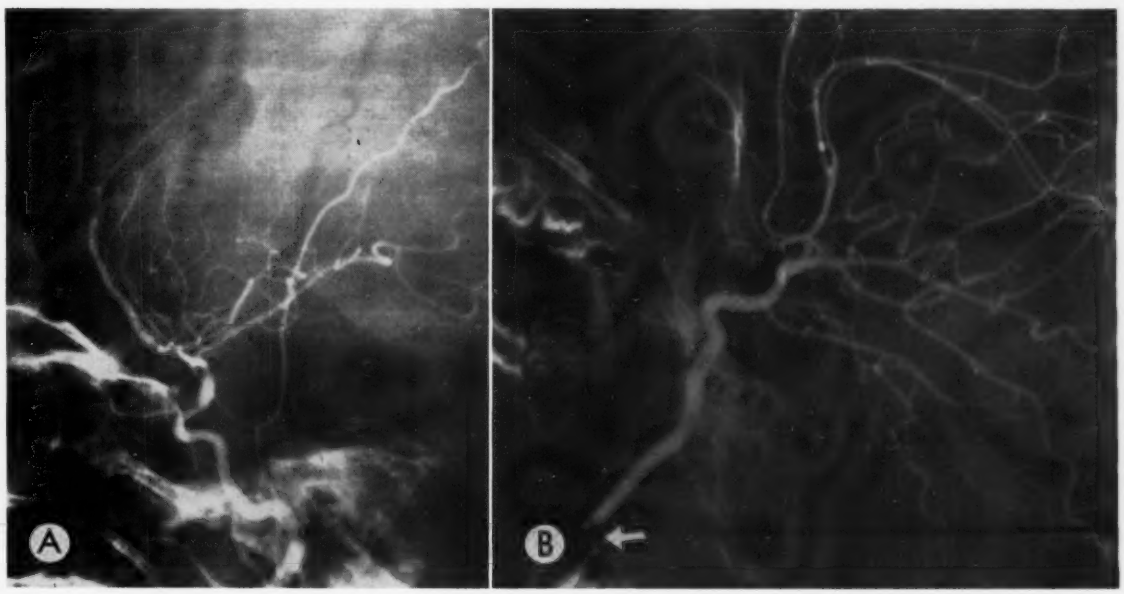



Fig. 17. (*A*) Local and diffuse spasm of cerebral arteries in a ten year old boy (unit no. 122-38-39) with persistent blood in the cerebrospinal fluid apparently secondary to operative injury. The history was that of severe head trauma followed by basal arachnoiditis and internal hydrocephalus, necessitating a short-circuiting procedure. There are several local areas of narrowing, undoubtedly due to spasm, in the carotid siphon and in the anterior and middle cerebral arteries noted both in lateral and in frontal serialograms. (*B*) Local spasm of the cervical portion of the internal carotid artery in a six month old infant. Above and below the narrow segment (arrow) the caliber of the internal carotid artery is normal and there is no change in the caliber of the intracranial vessels.

puncture. In one case where intracranial bleeding occurred secondary to operative injury caused by the insertion of a flexible catheter to perform a ventriculo-pleural shunt, local and more diffuse spasm was noted (Fig. 17*A*). In another case spasm in the cervical portion of the internal carotid artery, similar to that described in adults by Decker,[4] was noted (Fig. 17*B*).

There were 2 cases in this group in which an extraordinarily marked degree of diffuse arterial narrowing was present. In the first case a diagnosis of encephalitis was made and the patient recovered. The second case was of great interest because the arterial narrowing was only present on one side, the side on the larger ventricle (Fig. 18). It seemed to be due to a combination of increased intracranial pressure and stretching of the vessels. However, hydrocephalus *per se* does not produce such a pronounced degree of arterial narrowing (compare with Fig. 15). One wonders whether there may not have been an inflammatory process affecting the vessels in these 2 cases leading to diffuse spasm. The clinical diagnosis in the second case, made by exclusion, was Schilder's disease. The child died three months later but no autopsy was performed.

*Interesting Cases.* The following cases are of sufficient interest to be illustrated or mentioned in this paper. A case of diffuse sclerosis (probably Schilder's disease) was operated upon after an angiographic diagnosis of a temporal lobe mass because

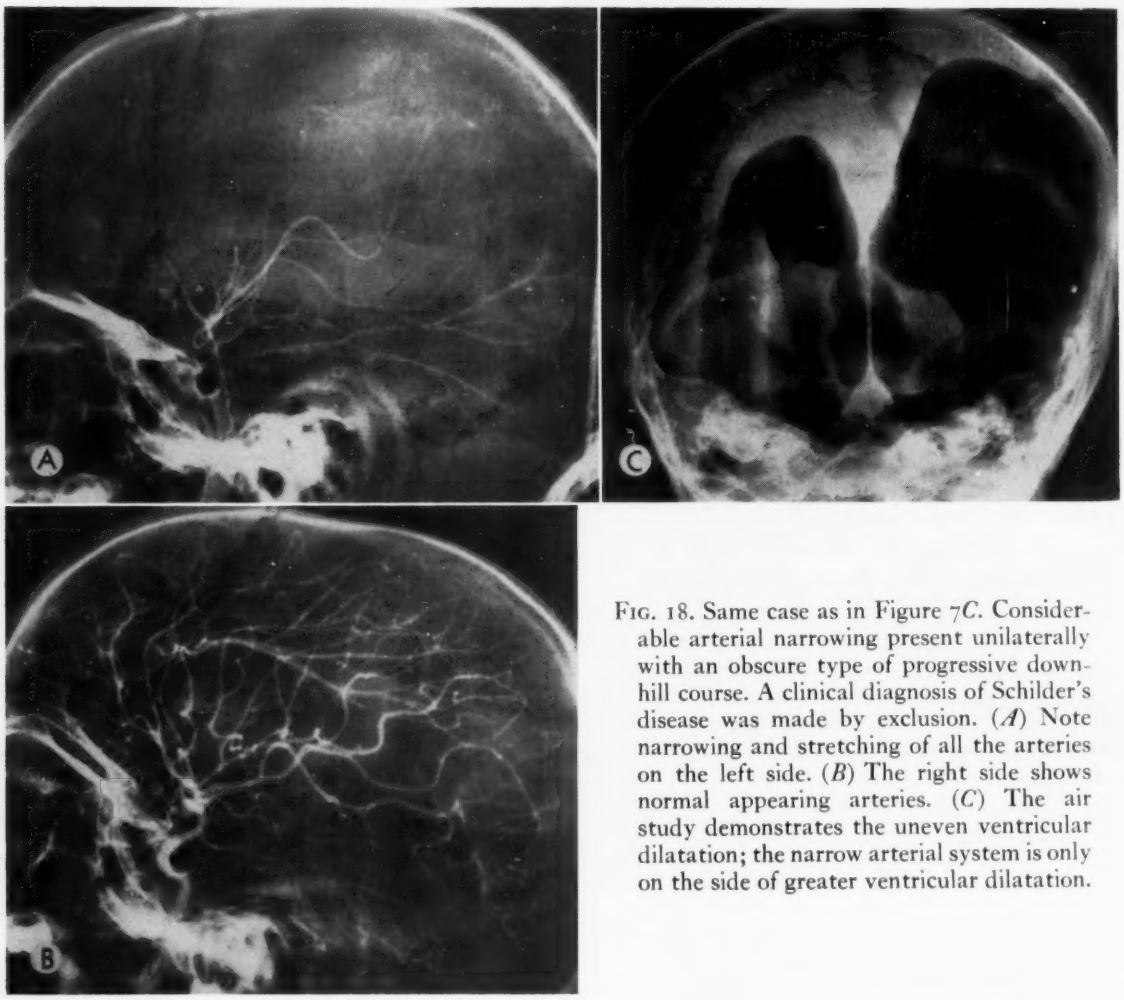



FIG. 18. Same case as in Figure 7*C*. Considerable arterial narrowing present unilaterally with an obscure type of progressive downhill course. A clinical diagnosis of Schilder's disease was made by exclusion. (*A*) Note narrowing and stretching of all the arteries on the left side. (*B*) The right side shows normal appearing arteries. (*C*) The air study demonstrates the uneven ventricular dilatation; the narrow arterial system is only on the side of greater ventricular dilatation.

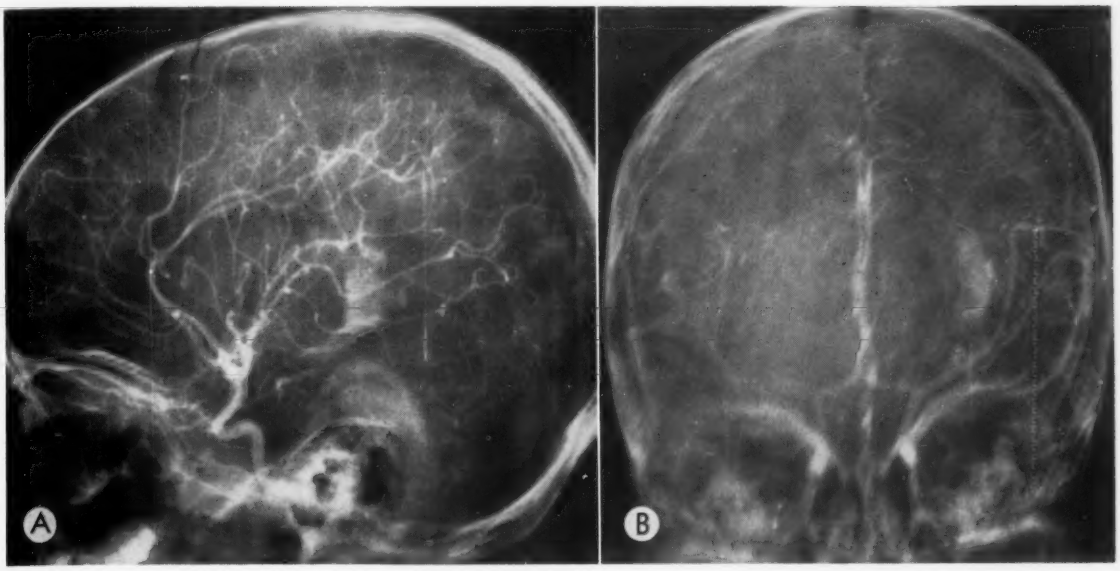



Fig. 19. Diffuse cerebral sclerosis (probably Schilder's disease) in a two year old child (unit no. 11-04-08) simulating a temporal lobe mass lesion. Note in *A* the elevation of the middle cerebral group. The middle cerebral branches have a "stiff" appearance as they come out of the sylvian fissure and over the temporal lobe. (*B*) The frontal angiogram confirms the elevation of the middle cerebral artery on the left side in comparison with the opposite side. At operation an abnormally hard temporal lobe was found. An incidental finding was that of extensive calcification in the glomus of the choroid plexus.

of an apparent elevation of the middle cerebral vessels (Fig. 19). At operation the temporal lobe was abnormal in appearance and firm. Another case had a very large porencephalic cavity, found to be under increased pressure, which produced angiographic signs compatible with a temporal lobe mass (Fig. 20). In an infant with hydranencephaly, angiography elucidated the pathologic anatomy of this congenital malformation. If hydranencephaly is merely an extremely marked degree of internal hydrocephalus, the cerebral vessels should be stretched over the surface sulci underneath the inner table of the skull. In our case the vessels were all near the base of the skull; the choroid plexus was rather prominently seen. This indicated a lack of development of the pallium rather than severe internal hydrocephalus.[13] In a similar case, clinically diagnosed as hydranencephaly (the skull transilluminated), the angiogram showed the middle cerebral branches over the surface of the brain, indicating that this patient had marked internal hydrocephalus.

SUMMARY

A study of 207 cerebral angiograms of children was made.

In taking these angiograms the technique was modified only to the extent that less angulation of the roentgen-ray tube was used in the anteroposterior projection than is customary in adults.

The following anatomic and physiologic features were found to be of interest from the angiographic standpoint:

1. The circulation time is, on the average, slightly shorter in children than in adults.

2. Filling of the posterior cerebral artery is more common than in adults.

3. The caliber of the intracranial branches of the internal carotid artery is definitely larger in children than in adults.

4. The middle cerebral artery is situated proportionally higher in children than in adults as related to a line that we have called the clinoparietal line.

5. The middle cerebral vessels move lower with advancing age, gradually approaching the adult configuration.

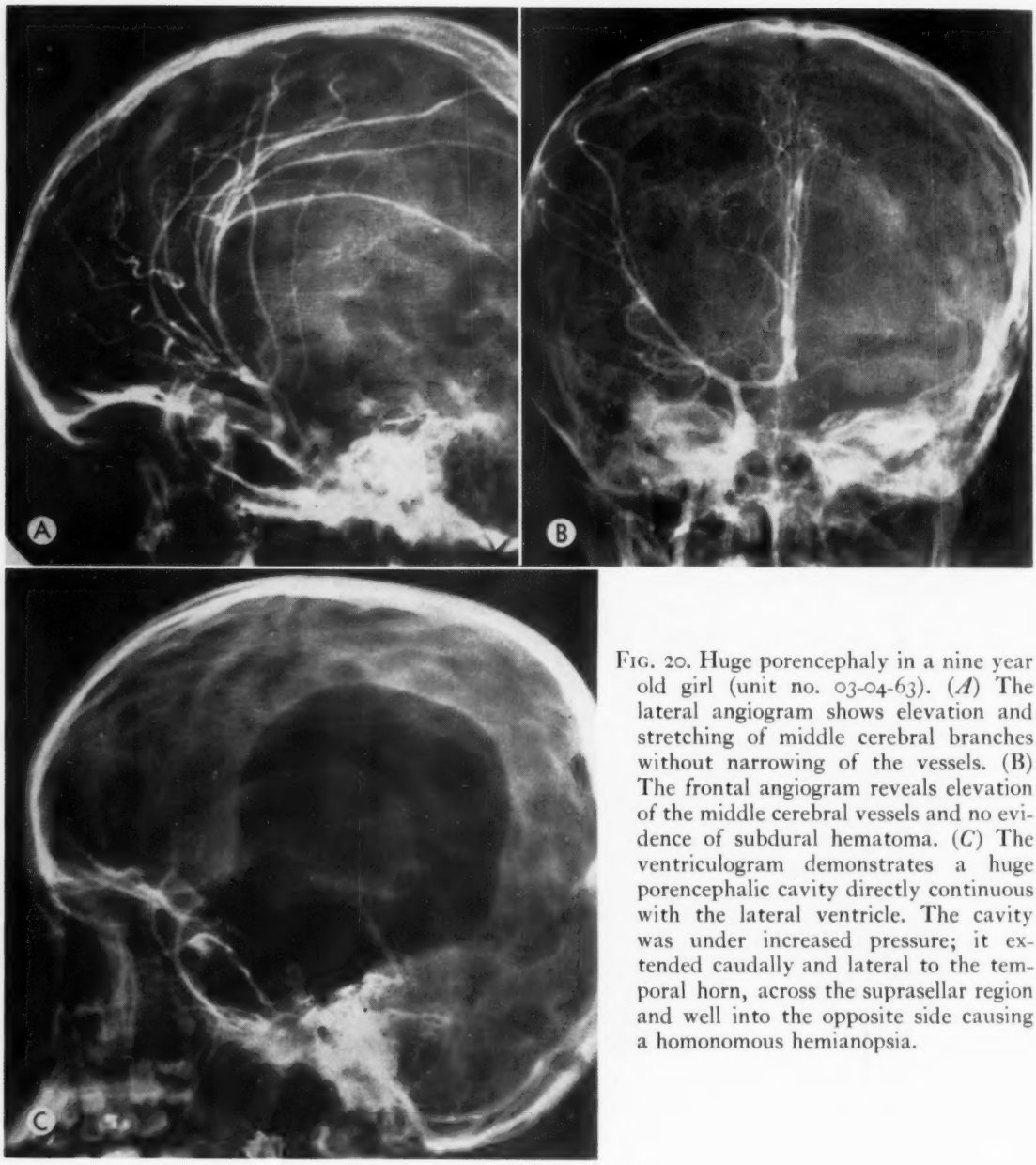



FIG. 20. Huge porencephaly in a nine year old girl (unit no. 03-04-63). (*A*) The lateral angiogram shows elevation and stretching of middle cerebral branches without narrowing of the vessels. (B) The frontal angiogram reveals elevation of the middle cerebral vessels and no evidence of subdural hematoma. (*C*) The ventriculogram demonstrates a huge porencephalic cavity directly continuous with the lateral ventricle. The cavity was under increased pressure; it extended caudally and lateral to the temporal horn, across the suprasellar region and well into the opposite side causing a homonomous hemianopsia.

Pathologically, one encounters the same conditions as in the adult, but the proportions are different. The incidence of arteriovenous malformations, subdural hematomas, and cerebral hemiatrophy in this group of children was higher than that encountered in adults in the same institution.

Juan M. Taveras, M.D.
Neurological Institute
622 West 168th Street
New York 32, New York

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# ABNORMAL CONGENITAL FISTULOUS COMMUNICATIONS OF THE CORONARY ARTERIES*

*By* GUIDO CURRARINO, M.D., FREDERIC N. SILVERMAN, M.D., *and* BENJAMIN H. LANDING, M.D.
CINCINNATI, OHIO

THE purpose of this paper is to report 3 instances of congenital fistulous communications between a coronary artery and a cardiac chamber, and to review this group of developmental anomalies of the coronary arteries.

These anomalies, even if rather uncommon, should be of special interest to the radiologist concerned with congenital heart diseases, as the diagnosis can at times be made by roentgen methods. Reviews of the subject have appeared recently in non-radiological journals.[20,56,60] The reader is referred to them for details not covered in this paper.

## REPORT OF CASES

CASE I. This white male was admitted to the hospital in 1954, at the age of eleven years, for evaluation of a cardiac murmur which had been present since birth. He had developed normally and had never shown any definite symptoms referable to the cardiovascular system. On physical examination, he appeared well developed and well nourished. The pulse rate was 90 per minute and water-hammer in character. The blood pressure was 110/50 in the arms and 140/60 in the legs. There was a left ventricular apical thrust and a grade 3 machinery murmur which had its maximal intensity at the fifth interspace in the right midclavicular line. At the cardiac apex and in the aortic area, there was a grade 2 systolic murmur. The rest of the physical examination was not remarkable. The electrocardiogram showed left ventricular hypertrophy. The roentgenograms of the chest showed slight cardiomegaly and mild pulmonary overcirculation (Fig. 1). A retrograde aortogram, with the tip of the catheter at the level of the aortic sinuses, revealed a grossly enlarged coronary artery which communicated freely with the lumen of the right ventricle (Fig. 2). As the patient was asymptomatic, no surgery was then advised. He was discharged on no special therapy or limitations and has remained entirely well since.

CASE II. This premature male infant was admitted to the hospital in 1947, at the age of ten days, for surgical correction of congenital atresia of the esophagus. The birth weight was 3 pounds, 15 ounces. On admission, there were clinical and roentgen signs of aspiration pneumonia (Fig. 3). A soft systolic murmur was heard over the precordium. Because of the lung disease, dehydration, and poor general condition, continuous suction to the upper esophageal pouch was instituted and only a gastrostomy was performed. In spite of these and other supportive measures, the patient's condition deteriorated rapidly and he died five days later at the age of fifteen days. At autopsy, in addition to the pneumonia and the esophageal

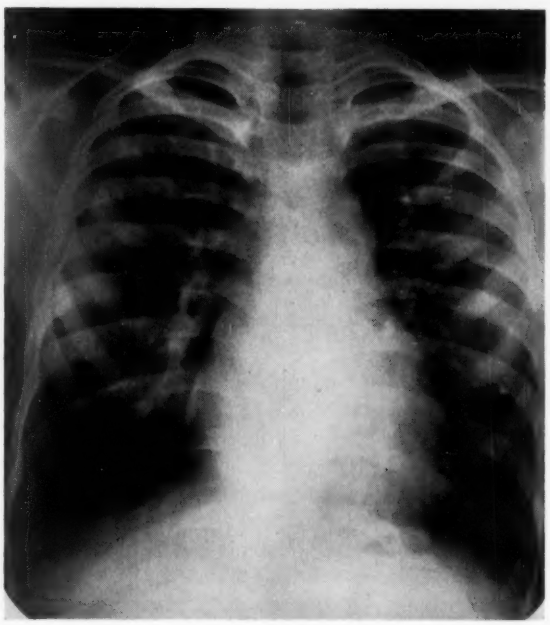

FIG. 1. Case I. Posteroanterior roentgenogram of the chest showing slight enlargement of the heart, main pulmonary artery and intrapulmonary vasculature.

* From the Departments of Radiology, Pediatrics, and Pathology, College of Medicine, University of Cincinnati, The Children's Hospital, and the Children's Hospital Research Foundation, Cincinnati, Ohio.

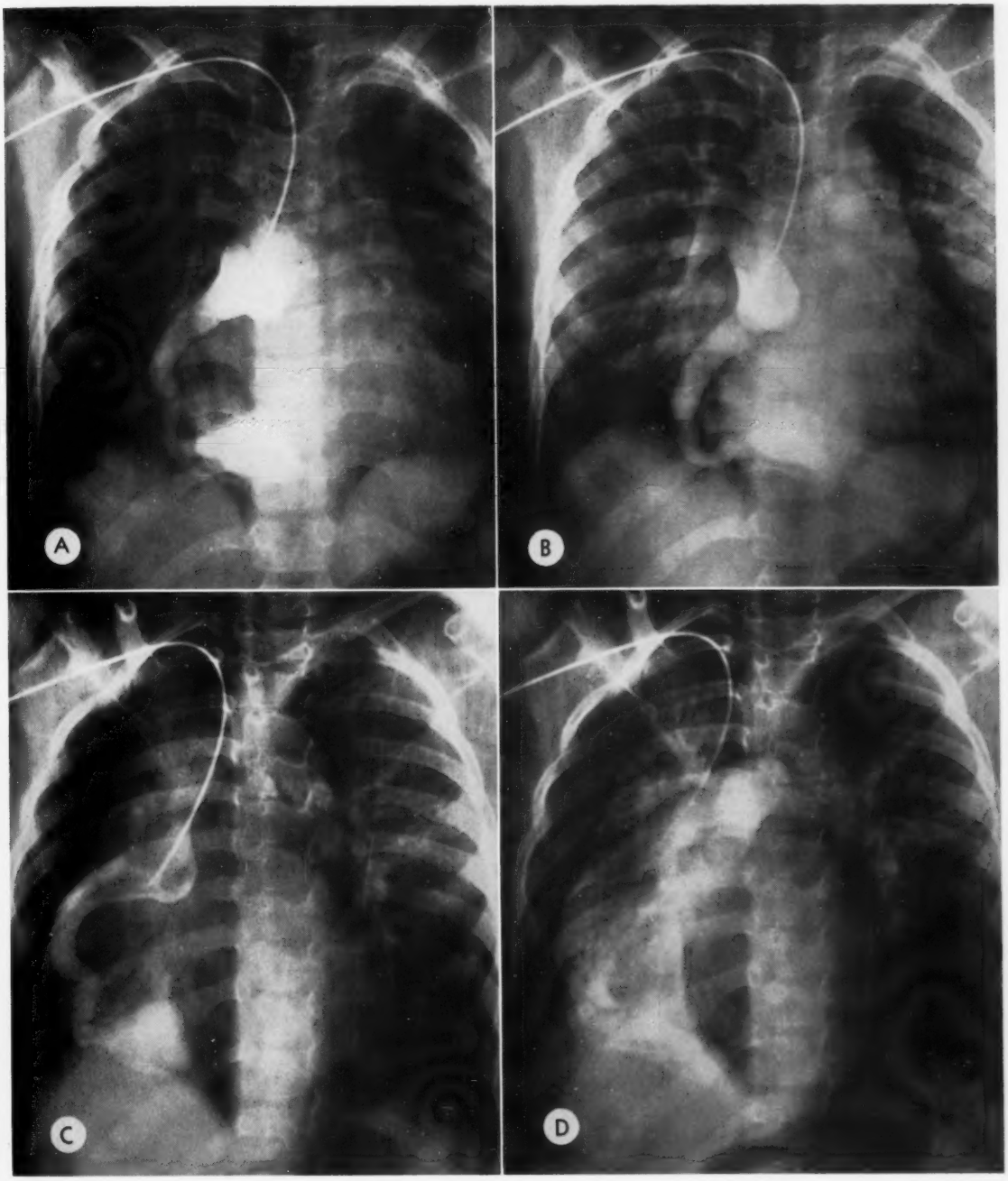


FIG. 2. Case 1. Retrograde aortogram with the tip of the catheter at the level of the aortic sinuses. Serial films exposed at a speed of four per second. (*A* and *B*) Anteroposterior projection. (*C* and *D*) Right posterior oblique projection. Note the enlarged and tortuous right coronary artery emptying freely into the right ventricle.

atresia (which was not associated with a tracheoesophageal fistula), a large supernumerary coronary artery was found arising from the posterior aortic sinus and emptying freely into the right atrium, just above the anterior leaflet of the tricuspid orifice. The other cardiovascular structures and the rest of the autopsy findings were not remarkable.

CASE III. This white female infant was admitted to the hospital in 1955, at the age of five days, for evaluation of cyanosis which had

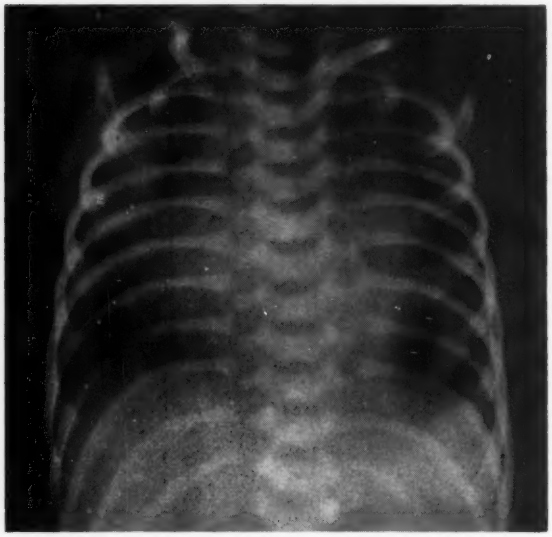

FIG. 3. Case II. Posteroanterior roentgenogram of the chest showing evidence of bilateral bronchopneumonia (aspiration) and slight cardiomegaly.

developed during the first twelve hours of life. She was born at term and her birth weight was 5 pounds, 13 ounces. On admission, she appeared well developed and well nourished. The cyanosis was moderately severe and generalized. The respiration rate was 50–60 per minute. The pulse rate was 160 per minute and regular. The femoral arterial pulses were normal. The eye lids appeared slightly edematous. The jugular veins were distended. The liver was two finger-breadths below the costal margin. There was a grade 2 systolic murmur, which had its maximal intensity to the left of the sternum, in the third and fourth intercostal spaces. The second pulmonic sound was single. An electrocardiogram showed marked right ventricular hypertrophy. The hemoglobin was 18.5 gm. per cent. Roentgenograms of the chest showed pulmonary undervascularity and a heart which was slightly enlarged and boot-shaped (Fig. 4). As the patient was considered to be in early cardiac failure, digitalis, mercurial diuretics and oxygen therapy were instituted. In spite of these measures, the patient's general condition gradually deteriorated. The liver became larger, and the respirations became labored and more rapid. The cyanosis became intense, especially when the patient was out of oxygen. A thoracotomy was performed under hypothermia on the seventeenth hospital day in an attempt to anastomose the left subclavian artery to the pulmonary artery, but was found to be unfeasible because of the small caliber of the pulmonary artery. The patient died five days later. The most important autopsy findings were limited to the cardiovascular system. The pulmonary orifice was atretic. The foramen ovale was open and there was a widely patent ductus arteriosus. On the anterior surface of the right ventricle there was an enlarged and tortuous right coronary artery, which communicated freely with the lumen of the right ventricle (Fig. 5). Microscopically, this vessel gradually lost its arterial muscular coat as it approached the lumen of the right ventricle. The sections of the myocardium showed enlarged embryonic sinusoids in the wall of the right ventricle. At several points these sinusoids communicated with branches of a coronary artery and with the right ventricle.

*Comment.* In the first patient, an eleven year old child, a fistulous communication between an enlarged right coronary artery and the right ventricle was demonstrated by retrograde aortography. The second case is that of an infant who, at autopsy, showed an enlarged supernumerary aortic coronary artery emptying freely into the right atrium. The autopsy on the third patient, also an infant, revealed a congenital atresia of the pulmonary artery orifice

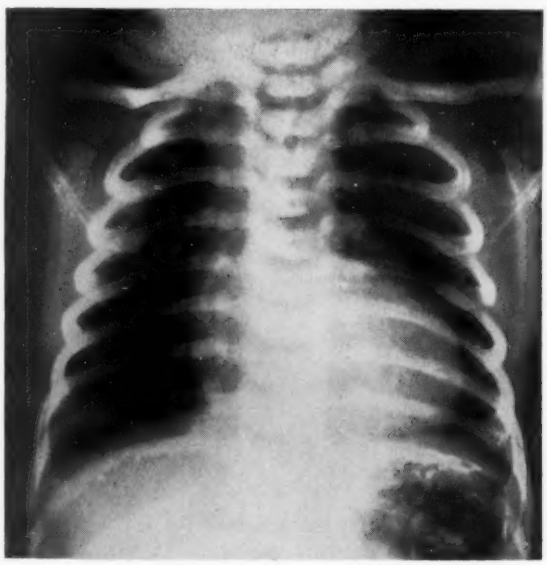

FIG. 4. Case III. Posteroanterior roentgenogram of the chest showing pulmonary undervascularity and a cardiac shadow which is slightly to moderately enlarged and boot-shaped.

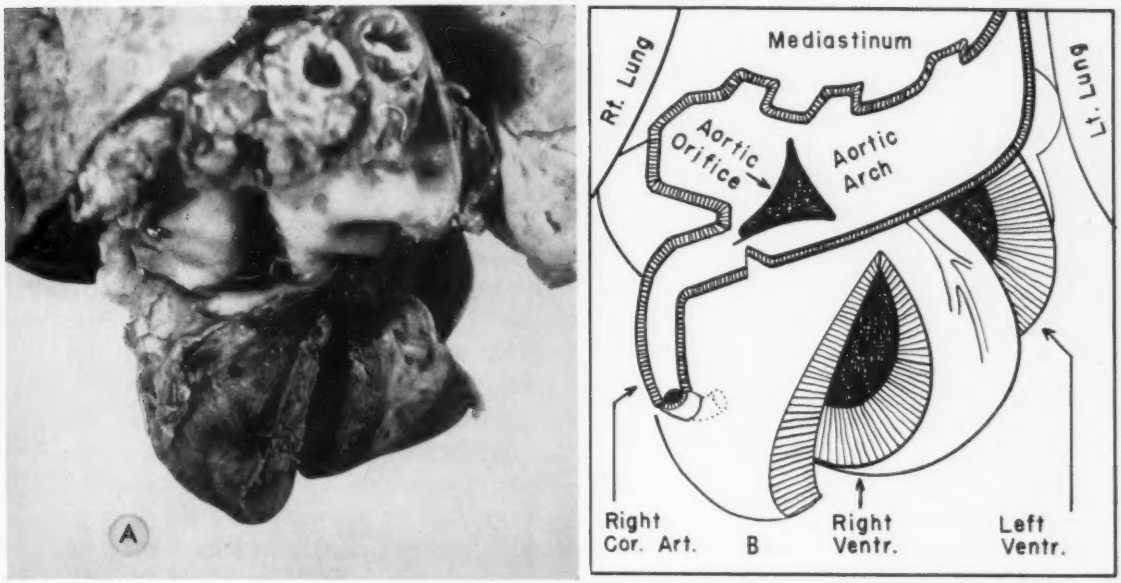


Fig. 5. Case III. (*A*) Photograph of the heart taken at postmortem examination. (*B*) Sketch of *A* showing an enlarged and elongated right coronary artery which empties into the right ventricle.

with intact ventricular septum and an enlarged right coronary artery which communicated freely with the lumen of the right ventricle. Only 2 cases with a similar combination of anomalies could be found in the literature.[2,62]

## DISCUSSION

### GENERAL CONSIDERATIONS

Congenital anomalies of the coronary arteries can be divided into anomalies of origin, number, size, and distribution; aneurysms; and abnormal communications. The abnormal communications consist of a gross fistulous connection between a main coronary artery, or one of its branches, and: (1) structures outside the heart, (2) the main pulmonary artery, (3) another coronary artery, (4) a coronary vein or the coronary sinus, and (5) a cardiac chamber. The coronary artery involved may be one of the two normal coronary arteries or a supernumerary coronary artery originating from the aorta or the pulmonary artery. The fistulous tract and the coronary artery from which it originates are often enlarged and tortuous and, occasionally, they are the site of large aneurysms. Microscopically, these anomalous channels may be arterial or partly arterial and partly venous. The size of their proximal and distal openings varies greatly from case to case.

The nature of these malformations is not certain, but since microscopic anastomotic channels normally connect the coronary arteries with all the structures mentioned above, it is possible that they are the result of dilatation of normal channels rather than anomalies which have developed *ex novo*.

The major communications of the coronary arteries are outlined in a diagrammatic form in Figure 6 and will be reviewed briefly. The coronary arterioles arborize into the vast myocardial capillary network from which arise the coronary veins; the coronary veins empty into the right atrium through the coronary sinus, with the exception of some anterior cardiac veins which empty directly into the right atrium. In addition to this circulation, there are numerous anastomotic channels between branches of the coronary arteries (interarterial anastomoses), between arteries and veins (arteriovenous anastomoses), and between branches of the coronary veins (intervenous anastomoses). The coronary

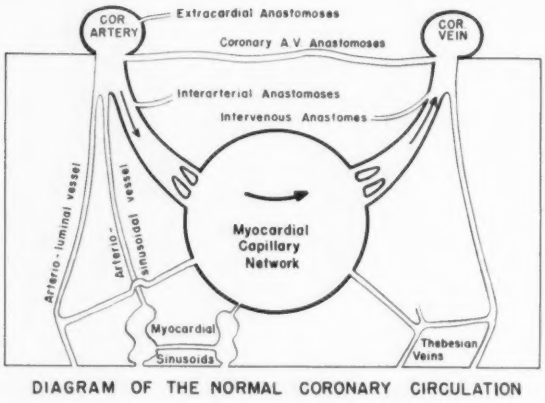


FIG. 6. The top of the diagram represents the epicardial surface; the bottom represents the endocardial surface. See text for details.

arteries also send anastomotic branches to arterioles of structures outside the heart (extracardiac anastomoses); these branches leave the heart from around the roots of the aorta, pulmonary artery, pulmonary veins, and venae cavae to be distributed to the vasa vasorum of these vessels, to the diaphragm, pericardium, mediastinum, esophagus, trachea, hili, bronchi, and the pleural surfaces of the lungs. The arterioles, with which these extracardiac branches of the coronary arteries anastomose, originate from the pericardiophrenic and other branches of the internal mammary artery, from the superior and inferior phrenic arteries, from anterior branches of the thoracic aorta, and from branches of the intercostal, pericardial, esophageal, and bronchial arteries. Numerous anastomotic channels also exist between the coronary system and the lumen of the heart. These channels include the arteriosinusoidal vessels, the arterioluminal vessels, and the thebesian veins. The myocardial sinusoids are simple endothelial tubes with regular lumina, which lie in intimate contact with the cardiac muscles; they represent the remnants of the intertrabecular spaces, through which blood flows directly from the lumen of the heart into the myocardium during early embryonic life, before the appearance of the coronary vessels. As the coronary veins and arteries grow, some sinusoids involute, whereas others decrease in size or become connected with the coronary arteries, thus giving origin to arteriosinusoidal vessels. The sinusoids anastomose freely among themselves and with the myocardial capillaries. The arteriosinusoidal vessels lose their muscular coat deep in the myocardium. The arterioluminal vessels are direct communications between branches of the coronary arteries and the cardiac chambers; they lose their muscular coat very near the endocardium. The thebesian veins are entirely venous channels which open directly into the lumen of the heart; they anastomose freely among themselves, with the myocardial capillaries and with branches of coronary veins. Further details and references on this subject may be found in works by Edwards[19] and Wearn.[61]

The cause for the development of the vascular malformations under discussion is unknown, especially in the majority of cases in which the lesion is not associated with other congenital cardiac defects. In those instances, such as Case III of the present report, in which the fistula communicates with a congenitally obstructed ventricle, it may be possible that increased pressure within this chamber in utero is responsible for the origin of the fistula by causing a dilatation of an arterioluminal or an arteriosinusoidal vessel. Further enlargement and elongation of the channel may be produced subsequently by the shunt itself, as it occurs in any other type of arteriovenous fistula throughout the body. Since the flow of blood during life in these grossly malformed hearts is probably from an obstructed ventricle toward the fistula, it is very likely that the anomaly acts as a useful decompressing channel.

## REVIEW OF THE LITERATURE

The normal communications of the coronary arteries are so numerous that it is not surprising to find so many types of coronary fistulae, if, for some reason, one of these normal anastomoses became enlarged. At least 52 instances of such malformations have been observed. All these cases are listed in four groups in Table I, with the ex-

| Type of Lesion | Author | Age* | Structures Involved in the Fistula | | Associated Cardiovascular Anomalies Remarks |
|---|---|---|---|---|---|
| 1. Extracardiac coronary fistula | Brooks[9] (Case 2), 1885 | Adult | Right coronary artery and supernumerary coronary artery from pulmonary artery | Mediastinal vessels | |
| | Voss[59] | 37 yr. | Both coronary arteries | Bronchial arteries | Pulmonary atresia |
| 2. Fistulous communication between a coronary artery and the pulmonary artery | Krause,[38] 1865 | 53 yr. | Both coronary arteries | Main pulmonary artery | |
| | Brooks[9] (Case 1), 1885 | ? | Both coronary arteries | Main pulmonary artery | |
| | Feriz[25] (Case 2), 1923 | 8 yr. | Left coronary artery | Main pulmonary artery | |
| | Biörck and Crafoord,[6] 1947 | 15 yr.† | Left coronary artery | Main pulmonary artery | Surgery |
| | Scott,[53] 1948 | 84 yr. | Supernumerary aortic coronary artery | Main pulmonary artery | |
| | Allanby *et al.*[3] (Case 6), 1950 | 7 yr. | Supernumerary aortic coronary artery | Main pulmonary artery | Pulmonary valve atresia; ventricular septal defect |
| | Baylis and Campbell,[4] 1952 | 76 yr. | Left coronary artery | Main pulmonary artery | |
| | Walther *et al.*[60] (Case 2), 1957 | 14 yr.† | Right coronary artery | Main pulmonary artery | Thoracotomy; catheterization |
| 3. Fistulous communication between a coronary artery and a coronary vein or the coronary sinus | Halpert,[30] 1930 | 54 yr. | Right coronary artery | Coronary sinus (vein?) | |
| | Nagayo and Takahashi,[45] 1932 | 72 yr. | Left coronary artery | Coronary sinus | |
| | Löwenheim,[41] 1932 | 62 yr. | Right coronary artery | A coronary vein | |
| | Emminger,[21] 1947 | 43 yr. | Right coronary artery | A coronary vein | |
| | Paul *et al.*,[46] 1949 | 9 yr.† | Right coronary artery | A coronary vein | Thoracotomy |
| | Davison *et al.*,[17] 1955 | 58 yr. | Left coronary artery | Coronary sinus | Catheterization |
| | Knoblich and Rawson,[37] 1956 | 55 yr. | Left coronary artery | A coronary vein | |
| | Walther *et al.*[60] (Case 1), 1957 | 8 yr.† | A coronary artery | A coronary vein | Catheterization |
| 4. Fistulous communication between a coronary artery and a cardiac chamber | Livingston,[39] 1883 | 4 mo. | Supernumerary arotic coronary artery | Right cardiac chamber(s) | |
| | Cayla,[12] 1885 | 10 da. | A coronary artery (supernumerary aortic?) | Right ventricle | |
| | Blakeway,[7] 1918 | 36 hr. | Left coronary artery | Left ventricle | Aortic valve atresia |
| | Schöndube,[52] 1922 | cow | Left coronary artery | Right ventricle | Accessory right ventricle |
| | Reid[49] 1922 | ox | Left coronary artery | Left ventricle | |
| | Jacobi and Heinrich,[32] 1933 | 18 mo. | Supernumerary aortic coronary artery | Right ventricle (infundibulum) | Coarctation of the aorta |
| | Joest[33] | calf | Left coronary artery | Right ventricle | |
| | Raschke,[48] | cow | Right coronary artery | Right ventricle | |
| | Rubli,[50] 1933 | calf | Left coronary artery | Right ventricle | |
| | Harris,[31] 1937 | 43 yr. | Right coronary artery | Right atrium | |
| | Valdivia *et al.*,[58] 1947 | 25 yr. | Right coronary artery | Right atrium | |
| | Brown and Burnett,[10] 1949 | 13 mo. | Supernumerary aortic coronary artery | Right ventricle (infundibulum) | |
| | Essenberg,[23] 1950 | fetus | Supernumerary aortic coronary artery | Right atrium | |
| | Espino Vela *et al.*,[22] 1951 | 16 yr. | Right coronary artery | Right ventricle | |
| | Williams *et al.*,[62] 1951 | 4 da. | Both coronary arteries | Right ventricle | Pulmonary valve atresia; enlarged sinusoids |
| | Alexander and Green,[2] 1952 | 36 hr. | Left coronary artery | Right ventricle | Pulmonary valve atresia; very small ventricular septal defect |
| | Colbeck and Shaw,[13] 1954 | 85 yr. | Right coronary artery | Right atrium | |
| | Sondergaard,[55] 1955 | 11 yr.† | Left coronary artery | Right ventricle | Patent ductus arteriosus; surgery |
| | Mozen,[44] 1956 | 10 yr.† | Left coronary artery | Left atrium | Surgery |
| | Johnson[34] | 1 yr.† | Right coronary artery | Right atrium | Surgery |
| | Davis *et al.*,[16] 1956 | 19 yr.† | Left coronary artery | Right ventricle (infundibulum) | Catheterization; surgery |
| | Garomella[26] | Adult | Left coronary artery | Right atrium | |
| | Marrow[43] | Adult† | Right coronary artery | Right atrium | Retrograde aortography; surgery |
| | Edwards *et al.*,[20] 1958 | 53 yr. | Right coronary artery | Right atrium | |
| | Gasul *et al.*,[27] Case 1[24] | 7 mo.† | Right coronary artery | Right ventricle | Catheterization; angiocardiography; surgery |
| | Case 2 | 3 yr.† | Right coronary artery | Right ventricle | Catheterization; retrograde aortography; surgery |
| | Case 3 | 9 yr.† | Right coronary artery | Right ventricle | Catheterization; retrograde aortography, angiocardiography; surgery |
| | Present Authors, Case I | 11 yr.† | Right coronary artery | Right ventricle | Retrograde aortography |
| | Case II | 15 da. | Supernumerary aortic coronary artery | Right atrium | |
| | Case III | 22 da. | Right coronary artery | Right ventricle | Pulmonary valve atresia; enlarged sinusoids |

* All humans except where indicated. † Alive at time of original report.

ception of the cases studied by Gross,[29] Cooley and Sloan,[14] Wood,[63] and Steinberg *et al.*,[56] as the anatomic details of the lesion present are insufficiently known. It is very likely, however, that these four instances belong either to the third or to the fourth category.

1. *Extracardiac Coronary Arterial Fistulae.* Examples of fistulae corresponding to the normal extracardiac coronary anastomoses have been described by Brooks and Voss (Table 1, section 1). The case described by Brooks[9] (Case 2) was an adult who showed an aneurysmal mass of thin-walled arteries situated around the main pulmonary artery, trachea and aortic arch. This agglomeration of vessels was supplied by large branches of the right coronary artery, by branches of a supernumerary coronary artery originating from the pulmonary artery, and by branches from the left subclavian artery. The case reported by Voss[59] was that of a thirty-seven year old man with congenital atresia of the pulmonary artery orifice. Adequate circulation to the lungs was maintained by the bronchial and the coronary arteries. These coronary branches pursued a tortuous course to be distributed to the lungs, where they anastomosed with the bronchial arteries.

2. *Fistulous Communication between a Coronary Artery and the Main Pulmonary Artery.* Eight such cases have been collected (Table 1, section 2). The ages of these patients ranged from seven to eighty-four years. The left coronary artery was involved in the fistula in 3 cases, the right in one, and both coronary arteries in 2. In the other 2 instances the affected coronary artery was a supernumerary artery arising from the aorta. In one instance, atresia of the pulmonary orifice and a ventricular septal defect were also present. In 2 patients (Biörck and Crafoord[6] and Walther *et al.*,[60] Case 2) the diagnosis was made during life. These fistulae are thought to represent either dilated extracardiac branches of a normal coronary artery communicating with the pulmonary artery, or supernumerary coronary arteries arising from the pulmonary artery and anastomosing, by way of dilated interarterial anastomoses, with a normal coronary artery. It appears that when there is an accessory coronary artery originating from the pulmonary artery, this vessel has the tendency to show gross anastomoses with the normal coronary arteries.

3. *Fistulous Communication between a Coronary Artery and a Coronary Vein or the Coronary Sinus.* These connections are thought to be dilated coronary arteriovenous anastomoses. The ages of the 8 reported cases included in this group (Table 1, section 3) ranged from eight to seventy-two years. In 2 of these patients, the diagnosis was made during life (Paul *et al.*,[46] and Walther *et al.*,[60] Case 1). The right and left coronary arteries were involved in the fistula with an approximately equal frequency. A coronary vein was involved in 5 cases, the coronary sinus in 3.

4. *Fistulous Communication between the Coronary Artery and a Cardiac Chamber.* These channels are thought to represent dilated arterioluminal or arteriosinusoidal vessels. This group includes 30 cases (Table 1, section 4). In 5 instances, the anomaly occurred in cattle, but whether these animals are more susceptible than others to this type of malformation is not known. The ages of the persons ranged from fetal life to eighty-five years. In 9 of them (Davis *et al.*,[16] and Gasul *et al.*,[27] Case 1, 2 and 3; Johnson,[34] Marrow,[43] Mozen,[44] Sondergaard,[55] and authors' Case 1), the diagnosis was made during life. In the majority of cases, one of the two normal coronary arteries was involved in the fistula. The right and left coronary artery were involved with an approximately equal frequency. In only one instance both coronary arteries were affected. In 5 or 6 cases, the anomalous vessel was a supernumerary coronary artery, which arose either from the non-coronary sinus of the aorta, or adjacent to another coronary artery. In more than half of the cases, the fistula communicated with the right ventricle (occasionally at the level

of its outflow tract), in 10 cases with the right atrium, in 2 with the left ventricle, and in one with the left atrium. In addition to the coronary fistula, one case showed a coarctation of the aorta and another patency of the ductus arteriosus. In 4, the cardiac chamber, with which the fistula communicated, was congenitally obstructed (pulmonary atresia, aortic atresia), and in at least 2 of them the wall of this chamber showed enlarged embryonic sinusoids.

As an addendum to this third group of coronary fistulae, it will be of interest to recall several reported cases in which there was an incomplete coronary fistulous tract or in which the abnormal coronary communication was microscopic. These cases are not included in the table but are mentioned here as probable incomplete forms of of the anomaly under discussion. Abbott[1] reported a case of a sixty year old woman with an anomalous origin of the left coronary artery from the pulmonary artery. The anomalous artery communicated with a large triangular venous sinus located in the wall of the left ventricle. Some myocardial vessels emptied in the floor of this sac. Schauder[51] found in the heart of an ox an enlarged branch of the left coronary artery, which emptied into a sac-like structure in the interventricular septum. This sac did not communicate with the lumen of the heart. Craig[15] reported a case of an infant with pulmonary atresia; an enlarged branch of the left coronary artery entered a sinusoidal pattern of vessels in the wall of the right ventricle, which also contained enlarged sinusoids. Grant[28] described the heart of a fourteen month old infant with single ventricle, pulmonary atresia, and patent ductus arteriosus. In the ventricular wall, there was an aneurysmal cavity, which microscopically communicated with the lumen of the ventricle, with myocardial capillaries, and with branches of coronary arteries and veins. Trevor[57] described an eleven year old child in whom the descending branch of the right coronary artery was aneurysmatic and opened freely into the cavity of the right ventricle. This anomaly was associated with great dilatation of the rest of the right coronary artery and infective endocarditis in the wall of the fistula. The fistula was thought to be congenital in origin but to have become complete only a few days prior to death. The case reported by Lovitt and Lutz,[40] that of a sixty-two year old man, showed on the epicardium of the left ventricle an aneurysm composed of numerous endothelial-lined cavities which communicated, by way of an endothelial-lined channel, directly with the lumen of the left ventricle. The aneurysm microscopically communicated with branches of the left anterior descending coronary artery. Dilated myocardial sinusoids in the wall of the left ventricle have also been described in the otherwise normal heart of a dog by Burchell,[11] in the heart of a twelve hour old infant with hypoplasia of the left side of the heart and aortic valve atresia by Bellet and Gouley,[5] and in cases of anomalous origin of the left coronary artery.[18,35,36,42,47,54] In several of these instances the dilated sinusoids communicated with coronary artery branches and /or the lumen of the left ventricle. In the hearts showing dilated sinusoids and/or an incomplete fistulous tract (or microscopic channels) in the wall of an obstructed ventricle, the anomaly was possibly secondary to this congenital obstruction, as discussed previously.

### CLINICAL REMARKS

These remarks will be limited to those cases in which the anomaly is not associated with, and grossly overshadowed by, a severe congenital malformation of the heart, such as pulmonic or aortic atresia. The former are the most frequent and better known types of congenital coronary fistula.

A precordial murmur is a very frequent finding; this murmur is often continuous but, on occasions, it is systolic or both systolic and diastolic. Its maximal intensity may be located in the pulmonic area, an interspace or more below, or to the right of the midline. A definite correlation be-

tween the location of the murmur and the type and location of the lesion is not apparent. When a continuous murmur is heard best in the pulmonic area, the lesion is clinically indistinguishable from patency of the ductus arteriosus. When the continuous murmur is in other locations, the anomaly may have to be differentiated from an aorticopulmonic septal defect; collateral bronchial circulation; arteriovenous fistula in the lungs, mediastinum or chest wall; and rupture of an aortic sinus. In rupture of an aortic sinus of Valsalva, the murmur is acquired at the time of the accident and the rupture is accompanied by severe chest pain and other well known cardiac manifestations.

The systolic blood pressure does not seem to be significantly altered by the anomaly. In a few instances, the pulse pressure was increased slightly.

The electrocardiogram is usually within normal limits. Nonspecific electrocardiographic changes have been reported in some patients, however. Roentgenographically, the cardiac silhouette is normal in many cases. Cardiomegaly, enlargement of the main pulmonary artery, prominence of the intrapulmonary vasculature, and "hilar dance" have been noted in some instances. The presence of a large coronary artery aneurysm may be suspected from the roentgenographic study or at fluoroscopy. As expected, cardiac catheterization is of limited value, as it is not possible by this method to distinguish the shunt produced by a coronary fistula from other types of shunt. An exception to this may be selective catheterization of the coronary veins through the coronary sinus in the presence of an arteriovenous fistula, as in the case reported by Walther *et al.*[60] (Case 1).

The anomaly can be demonstrated by angiocardiography, in its levocardiographic phase, and by retrograde aortography as in Case 1 of this report. This patient, and apparently also the 2 patients studied by Gasul *et al.*[27] and the patient observed by Marrow,[43] tolerated the procedure well; however, from these few instances alone, it is not possible to evaluate all the risks inherent with retrograde aortography in these malformations.

The prognosis is generally fairly good, in contradistinction to the prognosis in the case of fistula produced by the rupture of an aortic sinus of Valsalva. Many of the patients are asymptomatic. Cardiovascular symptoms such as exercise intolerance, or frank congestive failure, have been observed in some instances. Subacute bacterial endocarditis was mentioned in 3 cases.[29,32,57] Death could be attributed to the lesion only in a few subjects.

The lesion has been corrected surgically with disappearance or improvement of the signs and symptoms in 9 cases (Biörck and Crafoord,[6] Davis *et al.*,[16] Gasul *et al.*,[27] Case 1, 2 and 3; Johnson,[34] Marrow,[43] Mozen,[44] and Sondergaard[55]). In 3 additional instances (Gross,[29] Paul *et al.*,[46] and Walther *et al.*,[60] Case 2) a thoracotomy was performed but, for various reasons, no corrective procedure was undertaken. The operation consisted of ligation and sometimes also of division of the fistula, after the surgeon had made certain that no untoward effect resulted from temporary digital occlusion of the abnormal channel. For the time being at least, surgery probably should be recommended only in symptomatic patients; however, it is not beyond the realm of possibility that in the future surgery will be considered in all instances, as is now customary with patency of the ductus arteriosus.

## SUMMARY

Three new instances of congenital communications between a coronary artery and the lumen of the heart are described. In one case, the diagnosis was made by retrograde aortography, in the other 2 at autopsy. This group of developmental vascular malformations, the related literature, and the normal coronary circulation are briefly reviewed. The assumption is made that these abnormal communications represent failure of involution and enlarge-

ment of normally existing vascular structures.

Frederic N. Silverman, M.D.
The Children's Hospital
Elland and Bethesda Avenues
Cincinnati 29, Ohio

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# THORACIC AORTOGRAPHY VERSUS LEVO-ANGIOCARDIOGRAPHY

*By* PROF. A. CASTELLANOS,* DR. OTTO GARCIA,† DR. ELOINA GONZALEZ,‡ *and* DR. RAUL PEREIRAS§

HABANA, CUBA

MANY radiologists and cardiologists believe that levo-angiocardiography provides good visualization of the aorta and its branches and that, therefore, direct or retrograde thoracic aortography is not necessary. In our experience and in that of others, this was not found to be correct.

Although it is true that in some congenital or acquired lesions of the aorta levo-angiocardiography yields sufficient anatomic information, in other cases thoracic aortography will prove necessary, regardless of the technique employed. The aortographic method may be changed according to the need in the individual case, the aortic pathology, and the age of the patient.

When the radiopaque substance is injected via a peripheral vein, the right chambers of the heart and the pulmonary artery and its branches are the first to be opacified. In 1946, we called this phase of the contrast visualization "dextro-angiocardiogram." Then, the radiopaque substance leaves these vessels and is distributed throughout the pulmonary network, and later returns through the pulmonary veins to the left chambers of the heart, the aorta and its branches. We called this second phase the "levo-angiocardiogram."

If the aorta arises from the left ventricle, it has less roentgenographic density in the levo-angiocardiogram than the pulmonary artery, because the radiopaque substance has been diluted with the blood when it passed through the pulmonary capillary vessels. This reduced contrast of the left chambers of the heart and of the aorta can be improved by selective angiocardiography. With this method, contrast material is injected directly into the right cavities of the heart or into the main pulmonary artery, thereby reaching in a shorter period the left cardiac chambers. But even so, the opacification in the "levo" phase will always be less than that in the "dextro" phase.

Other factors may intervene in the same manner. The principal of these is the left to right shunt through a ventricular septal defect. In such cases, a variable amount of the contrast medium passes into the right heart and the pulmonary artery, causing a lesser amount to reach the aorta and, therefore, a lesser opacification of the aorta results. This, however, does not mean that the levo-angiocardiogram will be of little value in the diagnosis of aortic disease. On the contrary, in many cases the levo-angiocardiogram is sufficiently distinct to permit identification of a lesion of the aortic arch, the descending aorta, or of one of the great vessels of the aortic arch. In general, the greater the diameter of the aorta, the better will be the opacification.

In those pathologic cases in which the aorta over-rides the interventricular septum or when it completely arises from the right ventricle, its opacification will be very intense in the dextro-angiocardiogram. With direct injection of the aorta, either by the retrograde method or by a catheter introduced into a peripheral artery up to the aortic arch, the roentgenographic density of the aorta and its branches may be so marked as to render possible its distinction from the right and left cardiac chambers. For this reason, in many cases thoracic aortography provides anatomic details that the levo-angiocardio-

* Professor of Pediatrics, University of Habana, School of Medicine. Director of A. Castellanos Foundation for Medical Research.
† Director of the Research Department.
‡ Associate Medical Doctor.
§ Chief of the Radiological Department, Hospital Municipal de Infancia, Habana, Cuba.

gram can hardly provide and by its use proper diagnosis can be greatly facilitated. Special care must be taken, however, that the contrast material enter the left ventricle. This can occur in five different ways: (1) from the right to the left auricle; (2) from the right to the left ventricle through a ventricular septal defect; (3) directly from the right auricle to the left ventricle (ostium commune); (4) from the aorta to the left ventricle with a catheter introduced from a peripheral artery; and (5) by direct puncture of the left ventricle.

With adequate contrast, the left ventricle and the aorta are sufficiently visualized and levo-angiocardiography has the same value as thoracic aortography. Opacification of the left ventricle and aorta by injection through the catheter is used in the diagnosis of congenital cardiac lesions associated with hemodynamic investigations. This technique permits the visualization of different cardiac chambers in the same exploration. In such studies, contrast demonstration of the aorta is usually of secondary importance and not essential.

## DIFFERENCE BETWEEN INTRAVENOUS ANGIOCARDIOGRAPHY AND THORACIC AORTOGRAPHY

It is true that the performance of intravenous angiocardiography is easier and faster, while aortography carries certain risks regardless of the method used. Yet, it should be pointed out that, by using proper technique, aortography may be performed with very simple equipment and a small staff with comparative safety for the patient.

The mortality of thoracic aortography has also been exaggerated. As in cardiac catheterization, most of the accidents were reported when the procedure was new and experience lacking. In institutions with long experience, fatalities are practically nonexistent.

## THORACIC AORTOGRAPHY

The anatomic data provided by thoracic aortography depend essentially upon the technique used. At present, the techniques most frequently employed are the following: (1) via the left humeral artery (Castellanos-Pereiras,[13] 1939); (2) via the left common carotid artery (Freeman, 1948); and (3) via the catheter introduced into the radial, brachial or femoral artery (Radner,[22] 1948).

We are more familiar with the first two, but the former is particularly helpful in infants under one year of age, in whom we have obtained excellent aortograms by performing the injection manually. We described this procedure in 1938.

In older children we inject the left common carotid artery because it is larger than the humeral artery and allows the use of a large trocar. We do not have any experience in performing the injection through a catheter, although others have used this method with good results.

*Position of the patient.* The most advantageous positions for the diagnosis of patent ductus arteriosus and coarctation of the aorta are the left anterior oblique and the lateral. The roentgenographic details in the frontal position are very poor because of the superimposition of the structures; however, in this position a left subclavian artery arising directly from the aorta to the left side of the midline can best be diagnosed.

The frontal and lateral positions are very useful in the differential diagnosis of: aneurysm of the aortic arch and descending aorta; mediastinal tumors, depending on the localization of the tumor; aneurysms of the sinuses of Valsalva; and the incompetency or insufficiency of the aortic valves. In cases of vascular rings, the frontal position has given us good results.

*Seriographs and film changers.* In all our cases we have used a Fairchild magazine that permits the exposure of 2 films per second. Because of the injection time and the high speed of the blood in the aorta, generally only one film of diagnostic value is obtained. For example, the first film shows the arrival of the contrast medium in the ductus arteriosus, and the following

one shows it in the trunk of the pulmonary artery and its branches; hence, it is impossible to see the ductus arteriosus directly on the second film. We have reported elsewhere the procedure of compression of the lower part of the thoracic aorta in the epigastrium, an assistant pressing with his hand against the spine and upwards during the injection. This maneuver stops the blood flow in the aorta and the advance of the contrast medium is delayed a fraction of a second, thereby increasing the possibility of visualizing the ductus arteriosus.

Cinefluorography is undoubtedly the ideal method for thoracic aortography, because 50 frames per second can be exposed, but it is not essential. The rapid film or cassette changer enabling 4 to 6 exposures per second is usually quite adequate.

*Technique.* The best contrast media available at the present time are the 2-4-6 triiodo compounds which are better tolerated than the 3-5 diiodo compounds. In Cuba hypaque and renografin are the two media most commonly used.

With media of 70 or 90 per cent concentrations, 1.5 cc. per kg. of body weight is injected. By employing the retrograde route via a catheter through the left carotid or brachial artery, the entire amount can be injected in one second. At two seconds, thoracic aortograms of excellent quality are obtained, especially in adult or adolescent patients. As a rule, a concentration of 50 per cent is sufficient in newborns and small babies. In older children, a 70 per cent concentration produces a better contrast. Concentrations of 70 per cent or even 90 per cent (disodic salt of the 2-4-6 triiodo compounds) have been used through a No. 6 or 7 catheter in adults.

In infants under one year of age, the left humeral artery is preferred for injection of the contrast medium, as described in our original article in 1940.[13] If after dissection this artery proves to be too small and the surgical reconstruction after injection appears difficult, the left carotid artery is used. In all cases of coarctation of the aorta, the left humeral artery is selected because of the larger caliber of the neck and superior extremity arteries. Subject to the preference of the group performing the exploration, the injection is made by introducing the catheter into the radial (in coarctation), humeral or femoral arteries.

The trocar of choice is one manufactured by the Becton and Dickenson Company, No. 18 to 12. For cases in which the left common carotid artery is punctured, we reserve a large special model, No. 12, without a valve. The catheters used for injection are the Cournand or Lehman models No. 6 to 9, depending upon the age of the patient and the caliber of the arteries.

*Preparation of the patient.* Local anesthesia is used for injections into the humeral artery. A small amount of sodium seconal given intravenously makes it easy to place the patient in the correct position and suppress his movements. In adults, sodium pentothal is used when the carotid artery is injected. Occasionally, cyclopropane anesthesia may prove necessary.

After the procedure, the administration of an antibiotic and heparin within twenty-four hours is recommended.

## LEVO-ANGIOCARDIOGRAPHY

Three requisites are necessary to obtain good opacification of the left heart chambers and aorta in angiocardiography: (1) inject the contrast medium within one second; (2) inject an adequate amount of contrast medium; and (3) use an adequate concentration. This applies to both peripheral intravenous injection and selective angiocardiography.

For the intravenous injection, a trocar or a cannula as large as the vein can admit is required. However, the use of automatic injectors allows a rapid flow through a trocar of small caliber. The amount to be injected should be at least 1.5 cc. per kg. of body weight per second. A concentration of 70 per cent is preferred, but with the disodic triiodo compounds the ideal is a 90 per cent concentration.

In patients over six to eight years old, the use of a seriograph with a selector is

recommended. Two or three roentgenograms only are made in the dextro phase, and then four or six rapid exposures per second in the levo phase, according to the need.

When the technique in levo-angiocardiography is a good one and the seriograph is used, an intensely contrasted aortic image is obtained. For this reason, we do not believe that cine-angiocardiography has any advantage over levo-angiocardiography.

## MATERIAL

This paper is based on our experience with the use of intravenous angiocardiography and thoracic aortography at the A. Castellanos Foundation in the Children's Hospital and at the private office of Dr. R. Pereiras since 1939. The last 49 cases studied, which are listed in Table I, were chosen for special evaluation. Their distribution is shown in Table II. Many more patients have been examined during this period, but only those cases were selected in which thoracic aortography was performed for the purpose of comparison with levo-angiocardiography.

## CLINICAL APPLICATIONS

After a Blalock-Taussig or Pott's operation a failure may result due to closing of the anastomosis. It is, therefore, important to determine whether or not the anastomosis is functioning. Intravenous angiocardiography usually is inadequate for this purpose, as already stated in a paper published by us in 1953,[4] whereas thoracic aortography yields very satisfactory information. The left common carotid artery seems to be the best route for injection of the contrast medium but, when the operation is performed on the right side, it is best to use the homologous artery (Fig. 1).

### AORTIC INSUFFICIENCY OR INCOMPETENCY

In cases of regurgitation of the blood from the aorta into the left ventricle during diastole, this phenomenon can be well demonstrated in the living patient as well as in cadavers by thoracic aortography.[5] In cadavers, the injection of the contrast material must be performed rapidly, in less than one second. An injection carried out slowly may produce error. Normally, neither in the living patient nor in the cadaver does the contrast material flow into the left ventricle when the proper technique is used.

### ANOMALIES OF THE VESSELS ARISING FROM THE AORTIC ARCH

There are numerous anomalies of the vessels arising from the aortic arch. Of special interest is the left innominate artery with right common carotid and right subclavian arteries representing the counter-part of the normal pattern. This anomaly is demonstrated on the levo-angiocardiogram in the frontal view only if there is not an over-riding aorta, but, in the dextro-angiocardiogram, it is shown also in the presence of an over-riding aorta. Another anomaly is a right subclavian artery arising from the aorta to the left of the midline, usually associated with dysphagia lusoria. Although in one case of our series no dysphagia was present, the esophagram was typical; hence, contrast medium was injected via the right brachial artery against the blood flow and the anomaly was clearly demonstrated (Fig. 2). We have not found a published case report of a living patient with an arteriogram similar to that of our patient.

### VASCULAR RINGS

The levo-angiocardiogram permits good visualization, and in many instances enables identification of the structures forming the vascular rings. The left root of Arkin, found in many cases of vascular ring with a right-sided aorta, is very well seen in the frontal projection. Of course, thoracic aortography with proper technique, especially when exposures in the two projections are used, results in a better image. The double aortic arch in some cases may be diagnosed by levo-angiocardiography but in other cases thoracic

TABLE I

| Case No. | Age | Clinical Diagnosis | Operation | Autopsy | Angiocardiography | Catheterization | Aortography |
|---|---|---|---|---|---|---|---|
| 1 RGA | 12 yr. | Tetralogy of Fallot | Yes | No | No | No | Patent ductus arteriosus |
| 2 MLP | 4 yr. | Coarctation of the aorta | No | No | Defective | No | Coarctation of the aorta, adult type |
| 3 NCR | 3 mo. | Truncus arteriosus communis | No | No | Left anterior oblique, pseudotruncus arteriosus | No | Pseudotruncus arteriosus, defective |
| 4 RVB | 6 yr. | Arteriovenous aneurysm | No | No | Anomaly of the innominate artery | No | Anomaly of the innominate artery |
| 5 JLR | 6 yr. | Coarctation with total transposition | No | No | Total transposition. Coarctation? | Yes | Complete interruption of the aortic arch |
| 6 GDN | 3 yr. | Ductus arteriosus? Aortic septal defect? | No | No | Normal aorta | No | Aortic insufficiency. Coarctation? |
| 7 AN | 5 mo. | Coarctation of the aorta | Yes | Yes | Coarctation. No visible ductus arteriosus | No | Coarctation, adult type |
| 8 MR | 35 yr. | Coarctation of the aorta | Yes | No | No | No | Coarctation, adult type |
| 9 JSE | 6 yr. | Transposition? Tetralogy of Fallot | Yes | No | Transposition of the great vessels | No | Postoperative. Permeable anastomosis |
| 10 PM | 6 yr. | Tetralogy of Fallot | Yes | Yes | Tetralogy of Fallot | No | Postoperative, Closed anastomosis |
| 11 TM | 17 yr. | Coarctation of the aorta | Yes | No | Defective | No | Coarctation of the aorta |
| 12 OGU | 4 yr. | Ostium commune | No | Yes | No | Yes | Normal |
| 13 SRS | 3 mo. | Ductus arteriosus? Eisenmenger complex | No | Yes | Tetralogy of Fallot | No | Ductus arteriosus? |
| 14 JRL | 16 yr. | Tetralogy of Fallot | Yes | Yes | Defective | Yes | Aortic insufficiency |
| 15 LVA | 14 yr. | Interventricular septal defect | No | No | Interventricular septal defect? | No | Normal |
| 16 AC | 2 yr. | Truncus arteriosus communis | No | No | Truncus arteriosus communis | No | Truncus arteriosus communis |
| 17 EC | 9 yr. | Ductus arteriosus | Yes | No | Postoperative | No | Defective |
| 18 GF | 3 yr. | Ductus arteriosus | No | No | Ductus arteriosus | No | Patent ductus arteriosus |
| 19 JM | 8 yr. | Coarctation of the aorta | No | No | Coarctation, adult type | Yes | Coarctation, adult type |
| 20 GM | ? | Tetralogy of Fallot | Yes | No | No | No | Postoperative. Closure of the anastomosis |
| 21 YG | 3 yr. | Truncus arteriosus communis | No | Yes | Truncus arteriosus communis | No | Pseudotruncus arteriosus |
| 22 GPT | 8 yr. | Coarctation | Yes | No | Coarctation adult type | No | No |
| 23 LE | 20 mo. | Pseudotruncus arteriosus | No | No | Pseudotruncus arteriosus | No | Pseudotruncus arteriosus |
| 24 IZ | 5 mo. | Interventricular septal defect and ductus arteriosus | No | Yes | Interventricular septal defect and ductus arteriosus | No | Ductus arteriosus without hypertension |
| 25 PG | 2 yr. | Ductus arteriosus with pulmonary hypertension | Yes | No | Ductus arteriosus with pulmonary hypertension | Yes | Normal |
| 26 AF | 9 yr. | Ductus arteriosus with pulmonary hypertension | Yes | No | Ductus arteriosus with pulmonary hypertension | Yes | Ductus arteriosus with pulmonary hypertension |
| 27 VM | 9 yr. | Eisenmenger complex and ductus arteriosus | Yes | No | Eisenmenger complex | Yes | No ductus arteriosus |
| 28 AC | 1 mo. | Interventricular septal defect | No | No | Interventricular septal defect or ductus arteriosus | Yes | No ductus arteriosus |
| 29 MCH | 5 yr. | Interventricular septal defect and pulmonary stenosis | Yes | No | Interventricular septal defect and pulmonary stenosis | No | Normal |
| 30 RC | 3 yr. | Patent ductus arteriosus | Yes | No | No | No | Patent ductus arteriosus |
| 31 LH | 1 mo. | Coarctation of the aorta | Yes | No | Aortic arch not visible in the levo-angiocardiogram | No | Coarctation, infantile type |
| 32 MFM | 1 mo. | Coarctation of the aorta | Yes | Yes | No | No | Coarctation, infantile type |
| 33 RG | 2 yr. | Coarctation and interventricular septal defect | Yes | No | No | No | Coarctation, infantile type ' |
| 34 EMH | 7 yr. | Truncus arteriosus communis | No | No | Pseudotruncus arteriosus | No | Ture truncus arteriosus communis |
| 35 RR | 5 mo. | Truncus arteriosus communis | No | No | True truncus, interventricular septal defect | No | Pseudotruncus arteriosus |
| 36 BM | 20 da. | Trilocular heart. Pulmonary agenesia. Two cavas | No | Yes | Two cavas. Very large septal defects | No | Normal |
| 37 NSH | 17 yr. | Coarctation of the aorta | No | No | Coarctation of the aorta | No | Defective. Indirect sign |
| 38 MO | 13 yr. | Aortic or mitral insufficiency | No | No | No | No | Aortic insufficiency |
| 39 CT | 1 mo. | Ductus arteriosus without hypertension | Yes | Yes | No | Yes | Ductus arteriosus without hypertension |
| 40 CFG | 3 mo. | Coarctation of the aorta | Yes | Yes | No | No | Coarctation, infantile type |
| 41 JFC | 5 yr. | Coarctation of the aorta | Yes | No | Coarctation not visible in the levo-angiocardiogram | No | Coarctation, adult type |
| 42 GB | 6 mo. | Tetralogy of Fallot | Yes | No | No | No | Postoperative. Indirect sign of anastomosis |
| 43 LM | 11 mo. | Patent ductus arteriosus | No | No | Ductus arteriosus | Yes | Direct visualization of ductus arteriosus |
| 44 ARH | 2 yr. | Patent ductus arteriosus | No | No | Defective | Yes | Ductus arteriosus |
| 45 BC | 7 yr. | Ductus arteriosus and pulmonary hypertension | No | No | Pulmonary stenosis | Yes | Ductus arteriosus |
| 46 BG | 7 yr. | Tetralogy of Fallot and atypical ductus arteriosus | No | No | Tetralogy of Fallot and atypical ductus arteriosus | Yes | Atypical ductus arteriosus |
| 47 GDP | 6 mo. | Tetralogy of Fallot and wide ductus arteriosus | No | No | Tetralogy of Fallot and wide ductus arteriosus | Yes | Wide patent ductus arteriosus |
| 48 JG | 4 mo. | Anomalous right subclavian artery | No | No | No | No | Anomalous right subclavian artery |
| 49 GM | 9 yr. | Tetralogy of Fallot. Blalock-Taussig operation, right side | Yes | No | No | No | Anastomosis, quite closed |

TABLE II

| Pathology | No. of Cases |
|---|---|
| Truncus arteriosus communis | 4 |
| Patent ductus arteriosus with or without pulmonary hypertension | 10 |
| Coarctation of the aorta—infantile type | 4 |
| Coarctation of the aorta—adult type | 6 |
| Extreme tetralogy of Fallot with wide ductus arteriosus | 1 |
| Complete interruption of the aortic arch | 1 |
| Tetralogy of Fallot with atypical ductus arteriosus | 1 |
| Tetralogy of Fallot with right Blalock-Taussig operation | 2 |
| Tetralogy of Fallot with left Blalock-Taussig operation | 2 |
| Tetralogy of Fallot—anastomosis (end-to-end) of left subclavian to left pulmonary artery | 1 |
| Tetralogy of Fallot with Pott's operation | 1 |
| Aortic insufficiency | 3 |
| Partial persistency of the truncus arteriosus | 1 |
| Right subclavian artery arising from the left side | 1 |
| Normal aorta with or without other anomalies | 7 |
| Pseudotruncus arteriosus | 3 |
| Anomaly of innominate artery | 1 |

aortography is necessary for an accurate diagnosis.

### TRUNCUS ARTERIOSUS COMMUNIS AND PSEUDOTRUNCUS ARTERIOSUS

It is not necessary to stress the difference between truncus arteriosus communis and pseudotruncus arteriosus.[6] In the former, both pulmonary arteries, right and left, arise directly from the aorta; in the latter, the pulmonary arteries arise from a trunk and are generally short and narrow (Fig. 3).

In 6 of our cases doubt existed as to

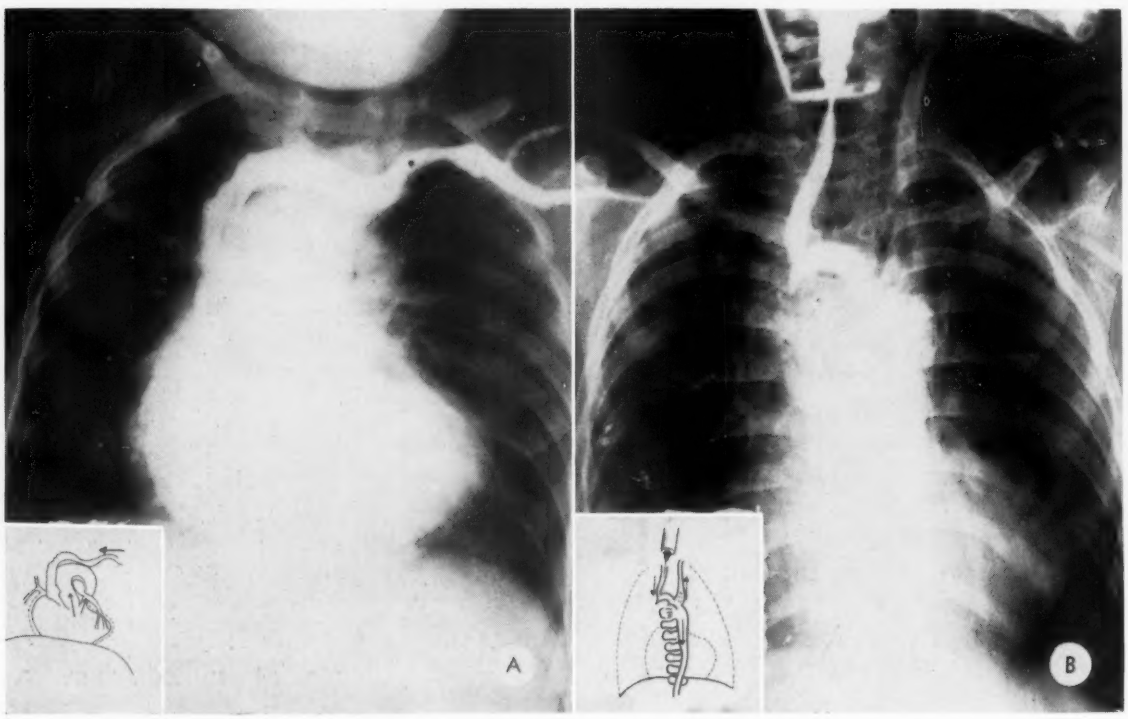


FIG. 1. Blalock-Taussig operation of right side. (*A*) Dextro-angiocardiogram; anteroposterior view. (*B*) Retrograde thoracic aortogram; anteroposterior view. The surgical anastomosis is closed.

whether a true truncus or a pseudotruncus arteriosus was present. Clinically, a true truncus arteriosus was diagnosed in 4; levo-angiocardiography and thoracic aortography confirmed the diagnosis. Of the remaining 2, one had a ductus of medium size and diameter, which was not visualized since only two exposures were made per second; in the other the frontal and lateral levo-angiocardiograms gave the impression of a true truncus arteriosus, but a thoracic aortogram demonstrated a ductus from which both pulmonary arteries arose.

Fig. 2. Retrograde arteriography via right brachial artery. Anteroposterior view. Anomalous course of the right subclavian artery.

### COARCTATION OF THE AORTA

Coarctation of the aorta is a congenital malformation which can best be diagnosed by thoracic aortography (Fig. 4), since more valuable information can be obtained from this procedure than from levo-angiocardiography (Fig. 5). A plain roentgenogram in the frontal position frequently shows in children the indentation or a "number 3 sign" which is of less signifi-

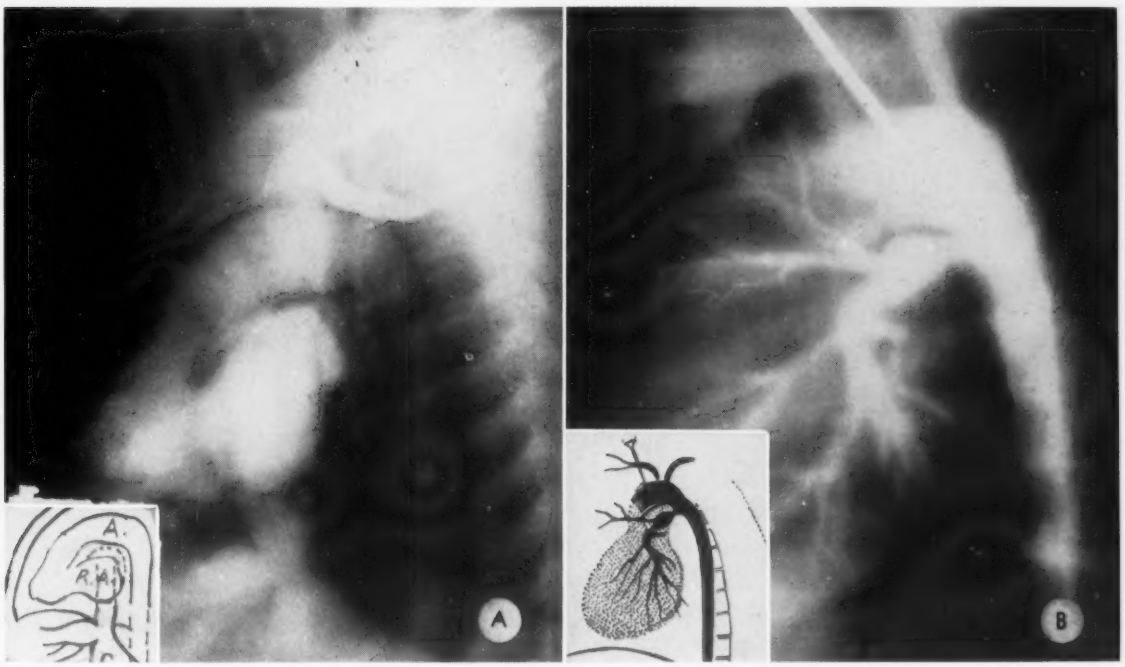


Fig. 3. (*A* and *B*) Pseudotruncus arteriosus; lateral views. The pulmonary arteries arise from a short narrow trunk.

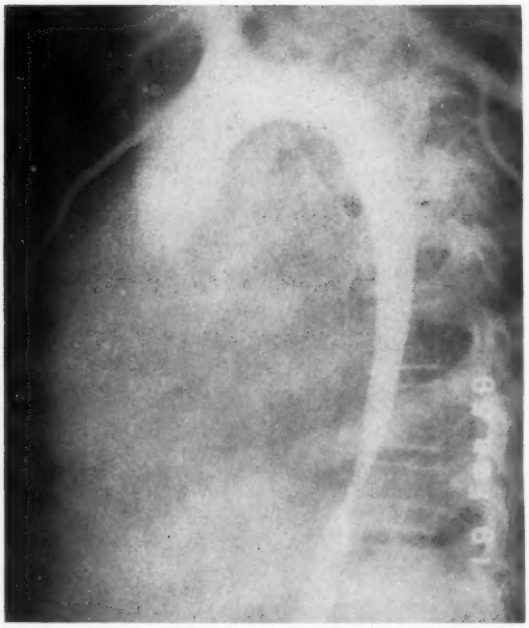

FIG. 4. Retrograde thoracic aortogram; left anterior oblique projection. Coarctation of the aorta and narrowing of the lower segment of the thoracic aorta.

cance than in the adult (due to the presence of the thymus gland). By using the standard technique of overpenetration in levo-angiocardiography, the coarctation can be demonstrated in the frontal projection, but in the left anterior oblique projection a better contrast can be obtained at the site of the narrowing.

When an atrial or ventricular septal defect coexists, the opacification of the aorta in the levo phase is so weak that it is of no diagnostic value in many cases (Fig. 6). The presence of a patent ductus arteriosus also contributes to poor contrast (Fig. 7). In some cases without ductus arteriosus or septal defects, in spite of the high concentration of the medium (70 per cent) injected within one second, and in spite of using the left anterior oblique projection, insufficient detail results. On the other hand, thoracic aortography in coarctation of the aorta (regardless of the patient's age, and the presence or absence of ductus arteriosus or septal defects) is always of great value for either diagnostic or surgical purposes.

Of 10 cases of coarctation of the aorta (4 of the infantile and 6 of the adult type), 5 patients had both intravenous angiocardiography and thoracic aortography and 5 had only thoracic aortography. Of the 10 thoracic aortographies, one (in a seventeen year old patient) was deficient because the injection was made via the brachial artery and the radiopaque substance was injected at a slow rate. The levo-angiocardiogram in this case demonstrated complete details of the narrowing. In the patients who had both procedures, the thoracic aortogram was greatly superior to the intravenous angiocardiogram. The roentgenograms of the patients who had only thoracic aortography were of excellent quality in 4. In one case with a defect of the aortic septum, the diagnosis could not be established because of the slow rate of the magazine.

### PATENT DUCTUS ARTERIOSUS

In this anomaly as well as in aortic septal defect, an ultra-rapid film changer is required in performing thoracic aortography. With only two exposures per second, there is not much possibility of obtaining direct visualization of the ductus arteriosus. The ideal procedure is cinefluorography, but by employing a fast seriograph enabling 4 or 6

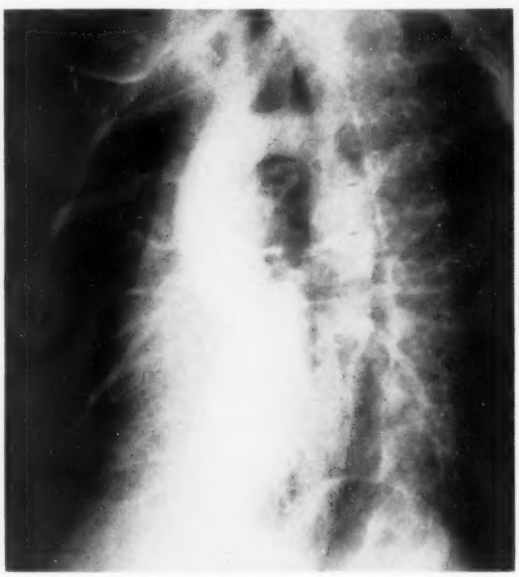

FIG. 5. Levo-angiocardiogram; left anterior oblique projection. Coarctation of the aorta, adult type.

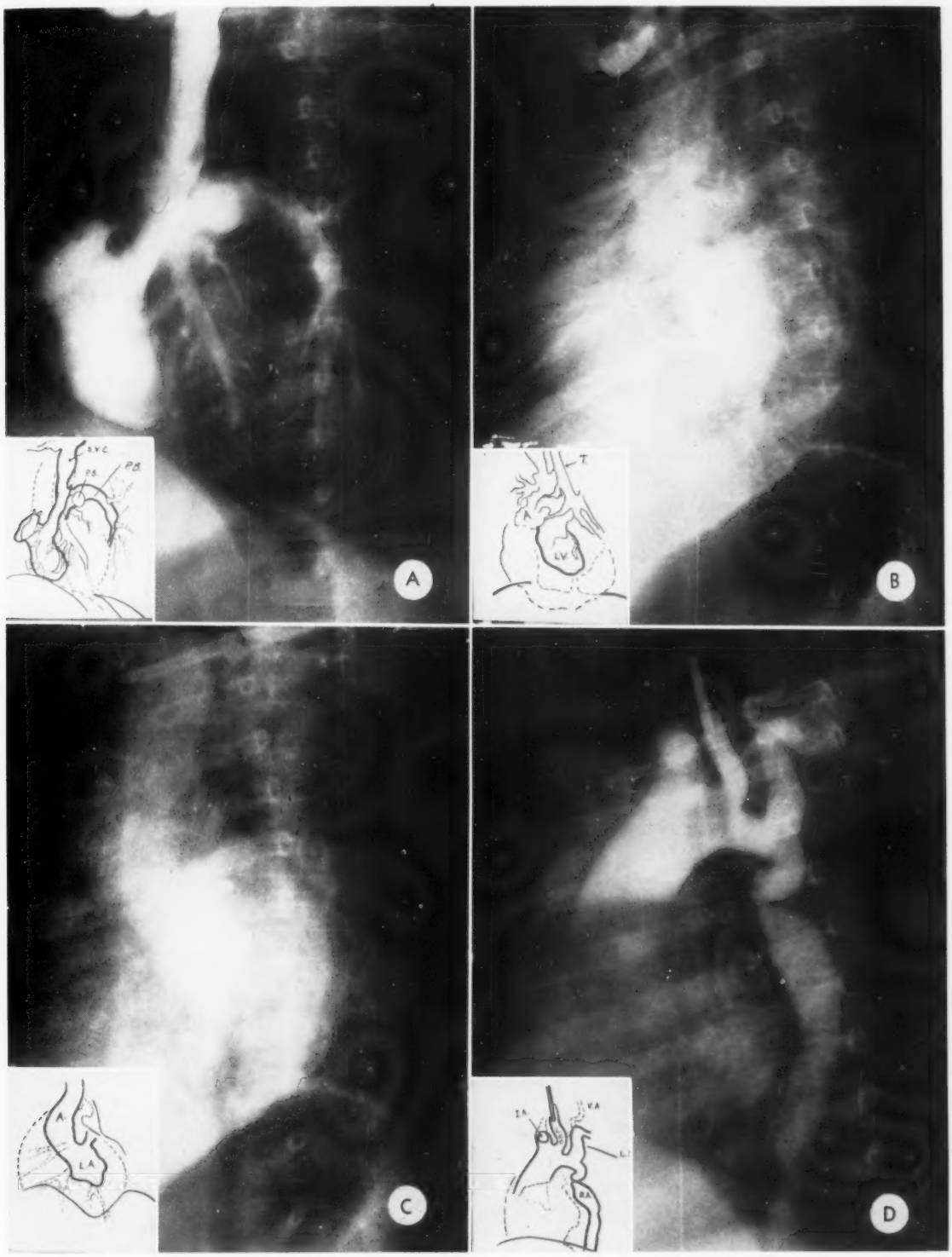


Fig. 6. (*A*) Dextro-angiocardiogram. (*B*) Levo-angiocardiogram. The aorta is not visualized. (*C*) The aortic arch is not clearly shown. (*D*) Retrograde thoracic aortogram. Coarctation of the aorta, adult type. The patient, five years old, also has an interventricular septal defect (catheterization).

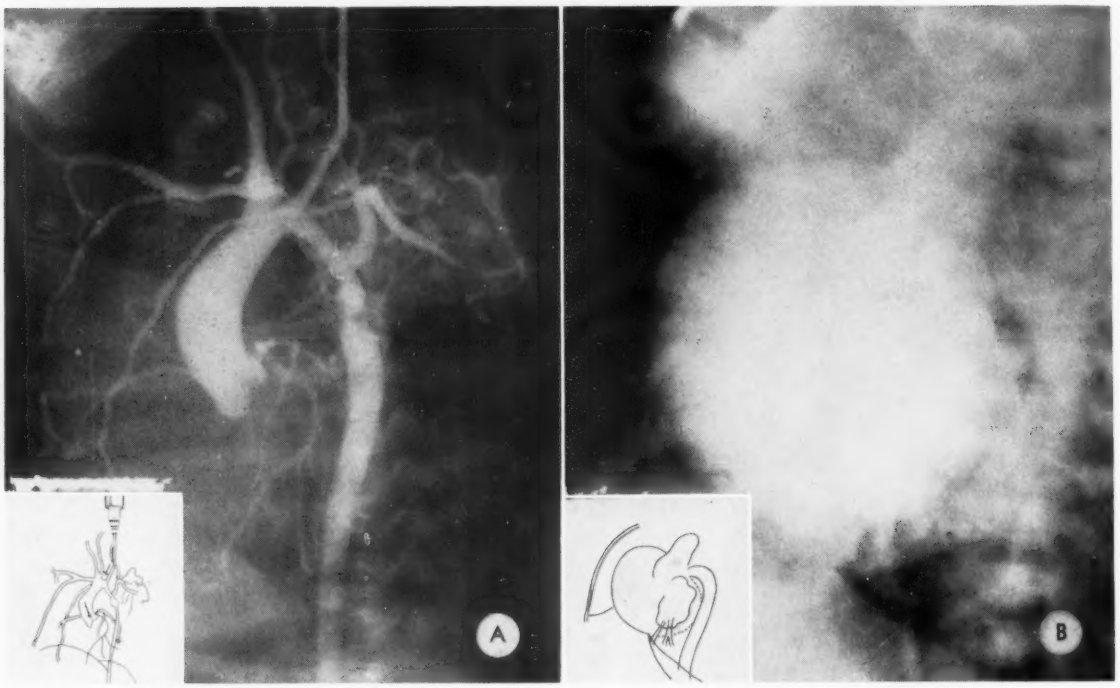


FIG. 7. (*A*) Retrograde thoracic aortogram. Coarctation of the aorta. (*B*) Levo-angiocardiogram; left anterior oblique projection. The coarctation is not clearly visualized. The patient, one month old, also has a permeable ductus arteriosus extending from the stenosis.

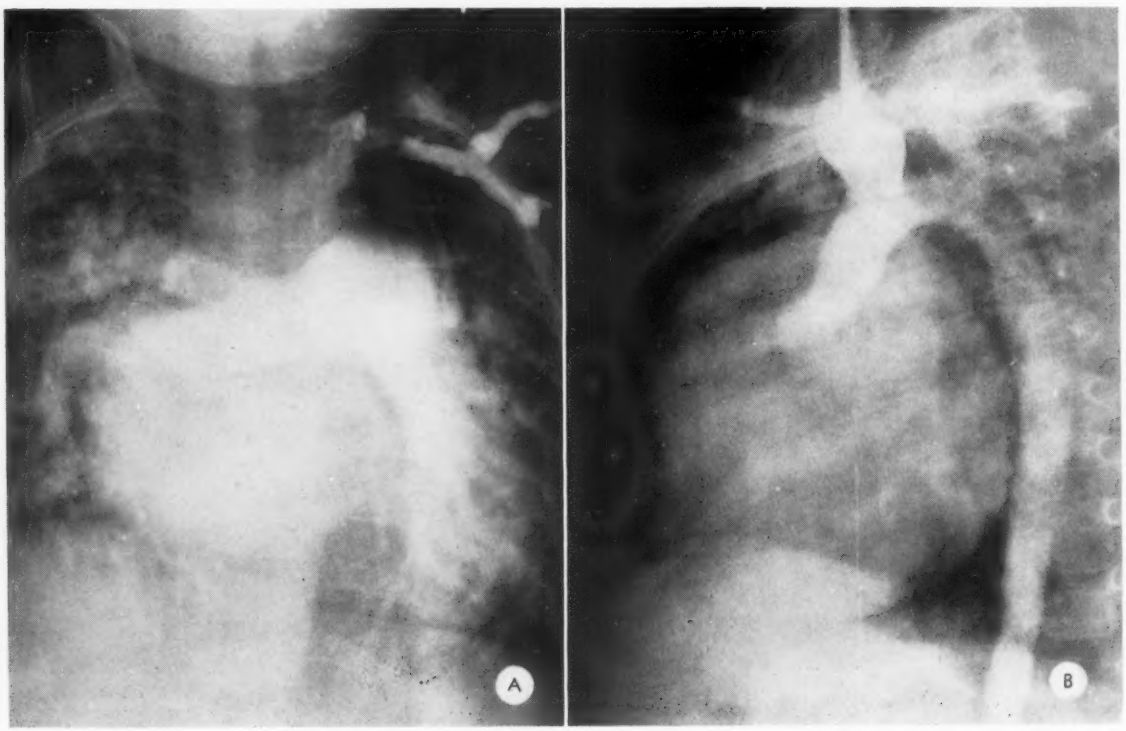


FIG. 8. (*A*) Dextro-angiocardiogram. Large pulmonary artery; slight opacification of the descending aorta. (*B*) Retrograde thoracic aortogram. Normal image. The patient has a patent ductus arteriosus and pulmonary hypertension.

exposures per second, the ductus arteriosus is always visualized in at least one film.

The results attained with classic intravenous angiocardiography and thoracic aortography depend on two factors: (1) the presence of other congenital cardiac lesions; and (2) the existence of pulmonary hypertension, and its severity. The most important of these is pulmonary hypertension in which dextro-angiocardiography is of greater value than levo-angiocardiography, especially when the pulmonary blood pressure is higher than the systemic blood pressure (Fig. 8). In the left anterior oblique or lateral projection it is possible to visualize the ductus arteriosus. However, when the pulmonary and the systemic blood pressures are equal and there is a wide ductus arteriosus with bi-directional shunt, re-opacification of the pulmonary artery may be obtained in the levo-angiocardiogram. When the systemic blood pressure is lower than the pulmonary blood pressure, thoracic aortography discloses that none of the contrast medium entered the main pulmonary artery.

In cases without pulmonary hypertension and left-to-right shunt, the direct visualization of the ductus arteriosus cannot always be attained with slow seriographs which are capable of only two exposures per second. In some instances the ductus arteriosus or the ductus arteriosus and main pulmonary artery may be seen in only one film. The maneuver of compressing the abdominal aorta, to which reference has already been made, in order to reduce the caliber of the descending aorta is not necessary in coarctation of the aorta but it is helpful in cases of patent ductus arteriosus without pulmonary hypertension, especially when a seriograph is used instead of cinefluorography.

We have performed thoracic aortography in a number of cases of patent ductus arteriosus (Fig. 9). However, we consider here only the atypical cases with systolic murmur and the cases with a very wide ductus arteriosus and cardiac insufficiency. In these cases catheterization

Fig. 9. Retrograde thoracic aortogram; left anterior oblique projection. Ductus arteriosus.

with the use of a contrast medium was necessary for an accurate diagnosis.

Of 10 patients in our series, 3 had pulmonary hypertension and 7 did not. Of the latter, the ductus arteriosus was totally visualized in 3, and partially visualized in 1. With proper technique and an ultrarapid film changer, it should be visualized in 100 per cent of the cases. The levo-angiocardiogram in the anteroposterior view permits a differential diagnosis between atypical ductus arteriosus and an anomalous coronary artery which terminates in the infundibulum. In both instances a continuous murmur is present.

The classic tetralogy of Fallot with a wide atypical ductus arteriosus and pseudotruncus arteriosus and a wide pulmonary artery was visualized in our series in the dextro-angiocardiogram only because of the over-riding of the aorta.

### Complete Interruption of the Aortic Arch

At least 12 cases of this anomaly have been reported recently; some were diagnosed during life. We have reported a case with transposition of the great vessels and two superior venae cavae, in which the diagnosis was made while the patient was

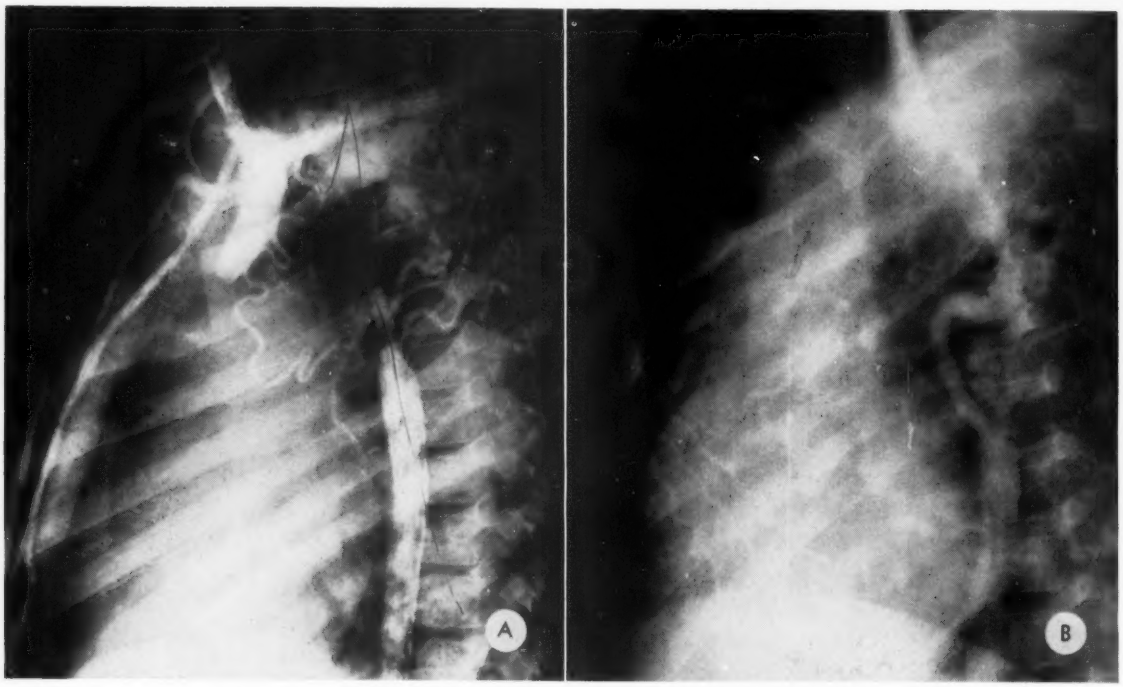


FIG. 10. (*A*) Retrograde thoracic aortogram. Complete interruption of the aortic arch. (*B*) One second later a large collateral vessel may be seen.

alive by one of us (Dr. O. Garcia). In another case, also diagnosed during life, surgical resection was tried unsuccessfully in 1958. The findings on thoracic aortography were typical. Roentgenograms were taken in the left anterior oblique projection and demonstrated innumerable collateral vessels which arose from the large arterial trunk originating in the right ventricle and ending in the distal part of the descending aorta (Fig. 10).

### ANEURYSM OF THE AORTA

In adults, frequently a distinction needs to be made between a solid tumor of the mediastinum and an aneurysm of the aortic arch or of the great vessels. Our experience on this subject is limited but we have seen cases in which intravenous angiocardiography has been of decisive value from the diagnostic point of view. The findings on the levo-angiocardiogram often were diagnostic but in very difficult cases direct opacification of the aorta is required for a more reliable diagnosis.

### ANOMALIES AND VARIATIONS OF THE CORONARY ARTERIES

Levo-angiocardiography is useless in the study of the coronary arteries. The anomalies and variation of caliber of the coronary arteries are best studied by thoracic aortography. It should be emphasized that in these cases injection of the contrast medium though the catheter is more advantageous than injection via the left common carotid or the brachial artery. However, congenital aneurysms of the coronary arteries may be easily diagnosed by levo-angiocardiography.

### CONCLUSIONS

1. Thoracic aortography is a procedure by which the thoracic aorta and its great vessels are best visualized since the contrast is sharper than that obtained in levo-angiocardiography.

2. In levo-angiocardiography the contrast of the thoracic aorta is often diminished because of the existence of a left to

right shunt through interatrial or interventricular septal defects.

3. In using intravenous angiocardiography, if the injection is not performed within one second or the concentration and the amount of the contrast medium are inadequate, the aortic opacification during the levo phase is much less than that in thoracic aortography.

4. The indications for thoracic aortography are numerous and comprise two groups: (1) those cases which require the use of levo-angiocardiography, and (2) congenital or acquired lesions which can only be diagnosed by this method.

5. Though many techniques have been proposed for thoracic aortography, three are used most often: (a) the retrograde route through the left humeral artery (Castellanos-Pereiras[13]); (b) the retrograde route through the left common carotid artery (Freeman); and (c) injection through a catheter placed into a peripheral artery (Fariñas,[15] Radner[22]).

6. The most suitable roentgenographic projection depends on the type of anomaly to be diagnosed.

7. Thoracic aortography is of greatest value in coarctation of the aorta, yielding considerably more information than can be obtained through levo-angiocardiography, especially in infants.

8. In patent ductus arteriosus, when studied with a seriograph, a fast device is required permitting at least 4 or 6 exposures per second; the use of cinefluorography is more advantageous.

Prof. A. Castellanos
23 No. 1.107
Vedado
Habana, Cuba

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VOL. 82, NO. 3

# ACYANOTIC TETRALOGY OF FALLOT*

*By* THEODORE E. KEATS, M.D., *and* JACK M. MARTT, M.D.
COLUMBIA, MISSOURI

WITH improved diagnostic acumen in congenital heart disease has come a better appreciation of the wide range of variability of the manifestations of any given type of congenital heart lesion. This has been proved true of the tetralogy of Fallot, which until recently has been thought to be fairly uniform in its clinical and roentgen manifestations. Our recent experience with 4 atypical cases of the tetralogy of Fallot, aged four, seven, thirty-seven and fifty-two, without cyanosis, and with unusual roentgen manifestations, as reported in this paper, serves to bring into even sharper focus the broad range of variation which may attend this condition.

## REVIEW OF LITERATURE

McCord and his co-workers,[2] in an excellent review of the tetralogy of Fallot, have called attention to the wide spectrum of clinical manifestations that is seen in patients with this anomaly, and have pointed out the variations in the degree of severity of underlying defects which lead to the different clinical pictures, including the absence of cyanosis.

Patients with the tetralogy of Fallot who are acyanotic include those with: (1) pulmonic stenosis of a mild or moderate degree accompanying a ventricular septal defect of small or moderate size. This combination of anatomic features frequently results in an evenly balanced pattern with one defect covering the hemodynamic effects of the other; (2) dominant ventricular septal defect with a mild infundibular stenosis. Physiologically these patients are characterized by a large left-to-right shunt at the ventricular level with an increased pulmonary blood flow; and (3) dominant pulmonic stenosis. This variation consists of a combination of severe right ventricular outflow obstruction and a small ventricular septal defect. The systolic pressure in the right ventricle may greatly exceed that in the left ventricle and a right-to-left shunt results, but is limited by the size of the defect.

Holladay and Witham,[1] in their discussion of tetralogy, have indicated the variability of its clinical manifestations including the acyanotic form. The over-all incidence of acyanotic tetralogy is said to be in the order of 10 per cent.[3]

## REPORT OF CASES

CASE 1. H. B., a thirty-seven year old service station attendant, entered the hospital complaining of dyspnea. A heart murmur was first detected at nine months of age. He had experienced shortness of breath with exertion for as long as he could remember. At the age of twenty-nine years he developed transient anterior chest pain which had recurred at irregular intervals. Dyspnea had become increasingly severe, forcing the patient to abandon employment. There was no history of cyanosis, squatting, or syncope.

Physical Examination. Blood pressure was 122/64 mm. Hg. No clubbing or cyanosis was noted. The heart was slightly enlarged in the transverse diameter and there was bulging of the anterior chest. A Grade IV harsh systolic murmur was audible at the third intercostal space at the left sternal border and was widely transmitted. Pulmonic closure sound was reduplicated.

An electrocardiogram revealed marked right ventricular hypertrophy. Hemoglobin was 14 gm. per cent.

Fluoroscopic examination of the chest showed no evidence of abnormal pulsations of the pulmonary arteries. Roentgenograms (Fig. 1, *A* and *B*) showed a normal heart size with evidence of hypertrophy of the right ventricle, dilatation of the right atrium and inferior vena cava, and abnormal tortuosity of the pulmonary vessels suggesting bronchial collateral circulation. An additional convexity on the left mid-cardiac border suggested an infundibular

* From the Departments of Radiology and Internal Medicine, University of Missouri School of Medicine, Columbia, Missouri.

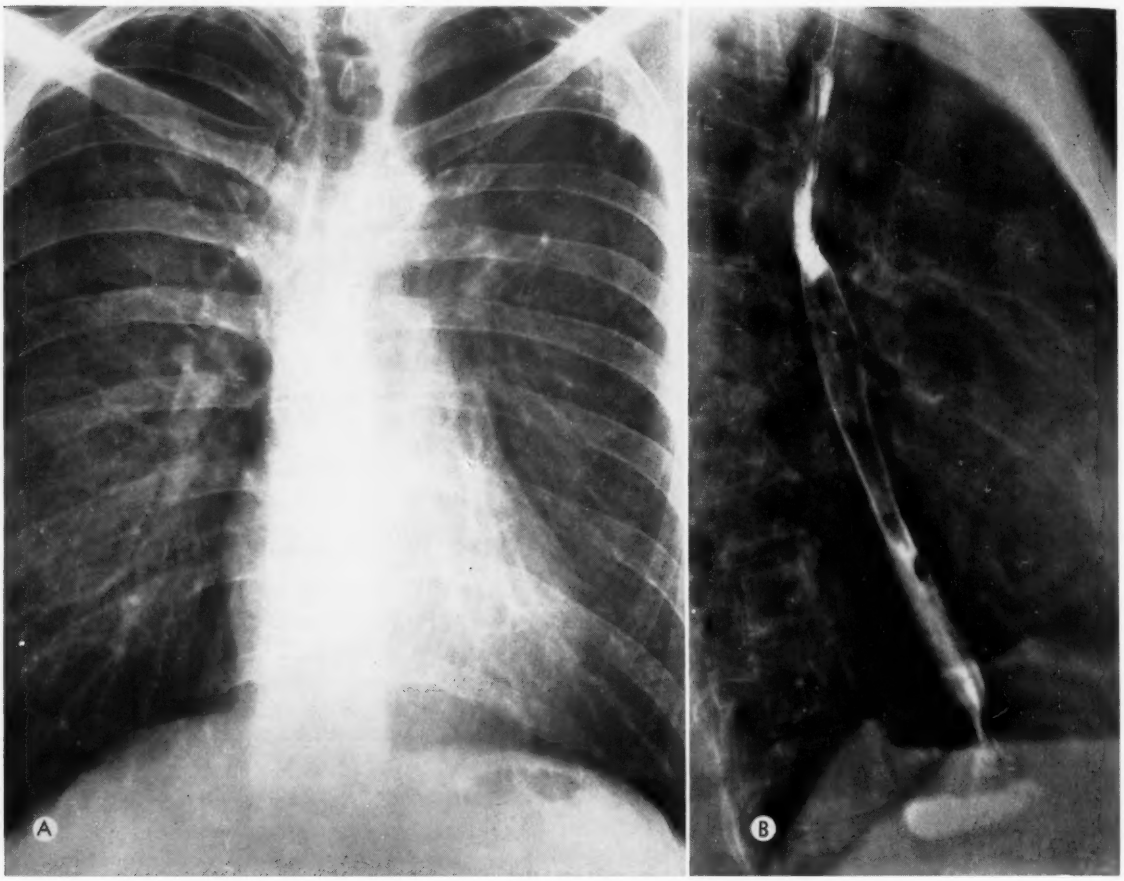


FIG. 1. (*A* and *B*) Posteroanterior and lateral roentgenograms show evidence of right ventricular hypertrophy, right atrial and inferior vena caval enlargement, bronchial collateral circulation, and convexity of the left mid-cardiac border indicating an infundibular chamber.

chamber. The left pulmonary artery was elevated by scarring resulting from an old left apical pulmonary tuberculosis. The roentgen impression was that of pulmonic stenosis.

Right heart catheterization revealed severe pulmonic stenosis and a ventricular septal defect, the catheter having entered the aorta from the right ventricle (Table 1). Arterial oxygen saturation was 86 per cent.

CASE II. R. E., a fifty-two year old gardener, was admitted because of dyspnea, fatigability, and ankle edema of progressive severity for one and one-half years. The patient was known to be "blue" at birth and was noted at intervals throughout his life to be slightly cyanotic upon heavy and strenuous exertion but not under ordinary conditions. He, nevertheless, worked daily and arduously. One and one-half years prior to admission he developed increased dyspnea, edema of the lower extremities, weakness, and fatigability, which were progressively severe. He received digitalis and diuretics, and although great improvement was noted, he did not recover sufficiently to return to work. There was no history of syncope, seizures, or squatting.

Physical Examination. Blood pressure was 140/90 mm. Hg. The mucous membranes of the mouth were questionably cyanotic. There was moderate anterior bulging of the chest wall. There was no clubbing of the fingers. The heart was markedly enlarged in its transverse diameter, the apical impulse being in the sixth intercostal space beyond the left anterior axillary line. A Grade IV harsh systolic murmur was audible in the fourth and fifth left intercostal spaces at the sternal border. The pulmonic closure sound was reduplicated but not accentuated. The liver was enlarged and tender. There was slight pitting edema of the ankles. Prominent "A" waves were observed in the jugular veins.

TABLE I

SUMMARY OF CATHETERIZATION DATA

| Case | Blood $O_2$ Content—Per Cent Saturation | | | | | Per Cent Shunt | Pressures mm. Hg | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | SVC | RA | RV | PA | Artery | | SVC | RA | RV | PA | Artery |
| I* | 71 | 76 | 75 | 75 | 86 | 26 R→L | 6/2 | 10/2 | 120/10 | 26/10 | 101/62 |
| II | 59 | 63 | 79 | 80 | 90 | 36 L→R | 10/4 | 9/4 | 117/12 | 31/19 | 135/75 |
| III* | 79 | 77 | 76 | 78 | 93 | 0 | 8/0 | 8/3 | 110/5 | 25/12 | 100/63 |
| IV | 73 | 73 | 85 | 85 | 99 | 53 L→R | — | 9/0 | 80/0 | 38/10 | 85/48 |

* Cathether entered aorta from right ventricle.

An electrocardiogram revealed incomplete right bundle branch block and right ventricular hypertrophy. Hemoglobin was 15.5 gm. per cent.

Fluoroscopic examination of the chest showed excessive pulsation of the right ventricle and its outflow tract. Intrinsic pulsations were noted in the pulmonary arteries. Roentgenograms (Fig. 2, *A* and *B*) showed generalized cardiac enlargement, right sided aortic arch and descending aorta, increased size of the main pulmonary artery and its branches and pulmonary hypervascularity. The roentgen findings were interpreted as evidence of a left-to-right shunt, probably an interventricular septal defect.

Right heart catheterization showed a moder-

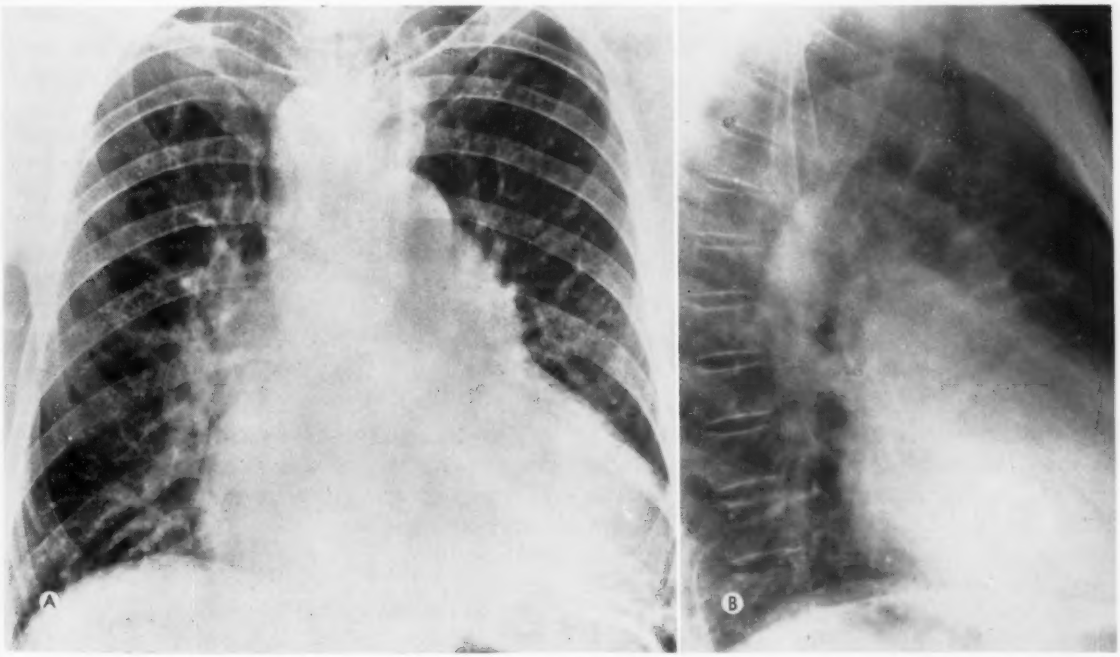


FIG. 2. (*A* and *B*) Posteroanterior and lateral roentgenograms show cardiac enlargement, right sided aortic arch with right descending aorta, prominence of the right ventricular outflow tract, and pulmonary hypervascularity.

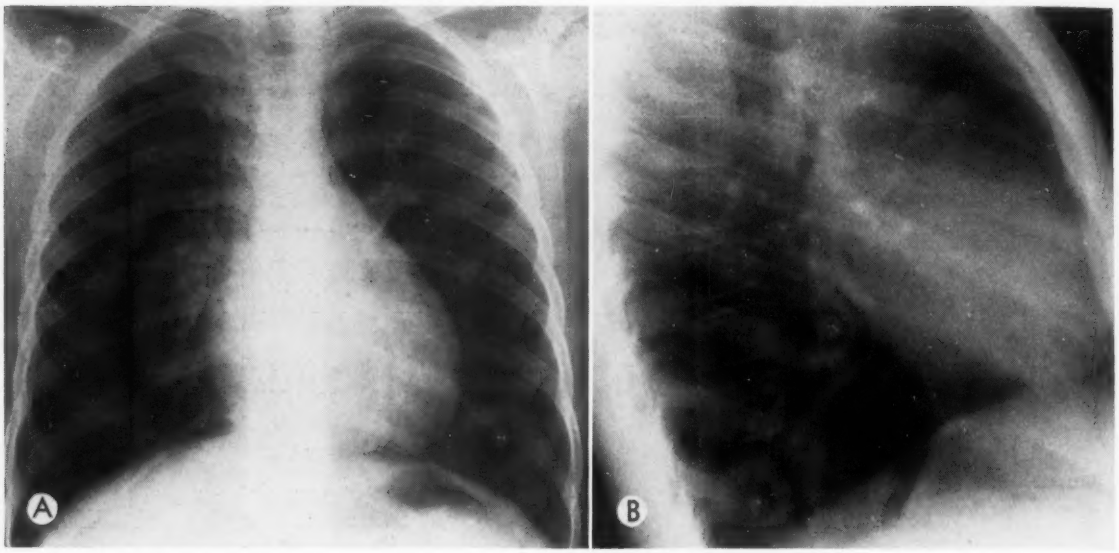


FIG. 3. (*A* and *B*) Posteroanterior and lateral roentgenograms show right ventricular hypertrophy, slight left atrial enlargement and normal pulmonary vascularity.

ately large left-to-right shunt at the ventricular level and severe pulmonic stenosis. Arterial oxygen saturation was 90 per cent (Table 1).

CASE III. A. O., a seven year old school girl, was admitted with complaints of shortness of breath and fatigability with exertion. On rare occasions she had been noted to squat after running. There was no history of cyanosis but a cardiac murmur had been noted at birth.

Physical Examination. Blood pressure was 110/63 mm. Hg. There was no cyanosis or clubbing of the fingers. There was slight increase in the transverse cardiac diameter. A Grade VI harsh systolic murmur was audible at the second left intercostal space and was associated with a marked systolic thrill. The pulmonic closure sound was inaudible.

The electrocardiogram was interpreted as right ventricular hypertrophy. Hemoglobin was 11.2 gm. per cent.

Roentgenograms of the chest (Fig. 3, *A* and *B*) showed no gross cardiac enlargement but did show evidence of hypertrophy of the right ventricle and slight enlargement of the left atrium. The pulmonary artery segment was not concave and the lungs showed normal vascularity. The roentgen impression was that of a small interventricular septal defect.

Right heart catheterization demonstrated an infundibular type of pulmonic stenosis and a ventricular septal defect as manifested by the cardiac catheter entering the aorta from the right ventricle (Table 1). Arterial oxygen saturation was 93 per cent.

CASE IV. E. B., a four year old girl, was referred because of easy fatigability and shortness of breath with exertion. Although cyanosis had not been observed by the parents, the child had on occasions squatted after running or playing. A cardiac murmur had been present since birth.

Physical Examination. Blood pressure was 84/55 mm. Hg. There was questionable cyanosis of the oral mucous membranes. The heart was moderately enlarged in the transverse diameter. A Grade V harsh systolic murmur was heard in the second and third left intercostal spaces at the sternal border and was associated with a systolic thrill. Pulmonic closure was slightly increased in intensity and closely reduplicated.

An electrocardiogram showed biventricular hypertrophy. Hemoglobin was 12.3 gm. per cent.

Roentgenograms of the chest (Fig. 4, *A* and *B*) showed generalized cardiac enlargement. There was slight prominence of the pulmonary artery and the lungs were hypervascular. The roentgen impression was interventricular septal defect.

Right heart catheterization revealed pulmonic stenosis and a ventricular septal defect with a moderately large left-to-right shunt at the ventricular level. The arterial oxygen saturation was 99 per cent (Table 1).

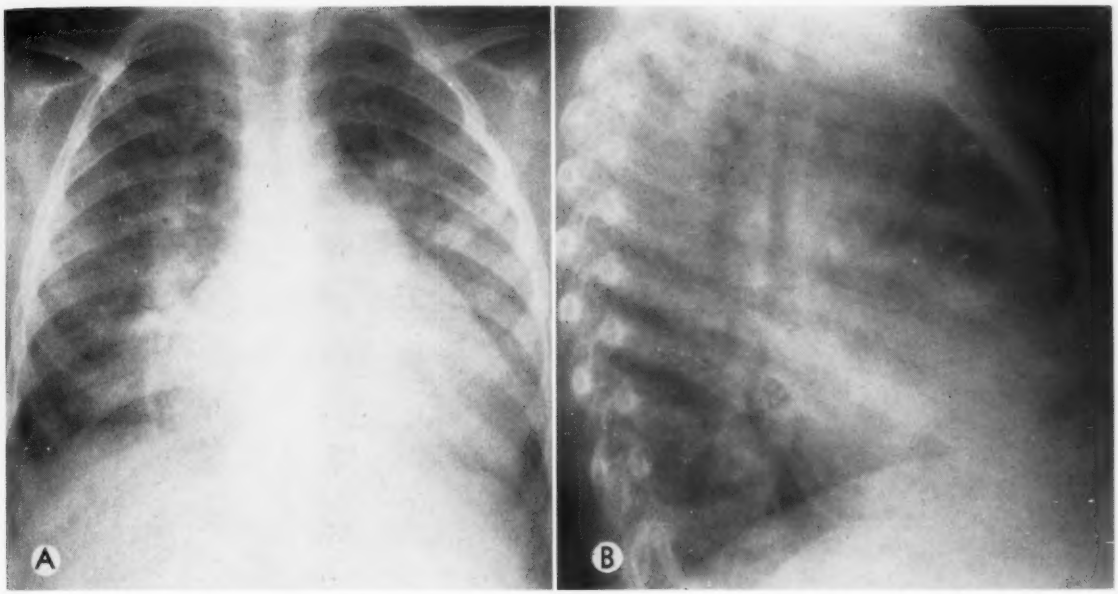


Fig. 4. (*A* and *B*) Posteroanterior and lateral roentgenograms show generalized cardiac enlargement, enlargement of the pulmonary artery and pulmonary hypervascularity.

COMMENT

The tetralogy of Fallot can now be demonstrated to occupy a broad spectrum in the field of congenital cardiac anomalies. While this entity has been termed a tetralogy, only two features, the pulmonic stenosis and the ventricular septal defect, are essential in the determination of the clinical and physiologic pattern of this defect.

Apparently, it is the size of the ventricular septal defect, the degree of pulmonic stenosis and the varying combinations of degree of these two defects which are responsible for the varying clinical and physiologic findings. The right ventricular hypertrophy is a secondary phenomenon. It is probable that the dextroposition of the aorta is of functional origin and not anatomic, relating to the defect in the membranous septum.

The "classic" appearance of the tetralogy of Fallot is well recognized and is illustrated in Figure 5. In its classic form the heart presents a blunted, uptilted apex, concavity of the pulmonary segment and hypovascular lung fields. However, the variability of the roentgen findings in this anomaly is amply demonstrated by the 4 cases reported here. The findings will, of course, correspond with the severity of the underlying defects.

Catheterization data obtained in Cases II and IV show a substantial left-to-right shunt at the ventricular level and, were it not for the pressure gradient across the pulmonic valve, the findings would be representative of an isolated large ventricular septal defect. The roentgen findings in these 2 patients likewise were interpreted as representing ventricular septal defects. In those cases of tetralogy of Fallot where the left-to-right shunt is a predominant element, the cardiac silhouette and the pulmonary vasculature will, of course, reflect the left-to-right shunt with enlargement of the pulmonary arteries and pulmonary hypervascularity. In these cases it is apparent, therefore, that the effect of the ventricular septal defect greatly outweighs that of the pulmonic stenosis. Consequently, minimal or no right-to-left shunting occurs which in turn accounts for the clinical absence of cyanosis.

On the other hand Cases I and III are noted to have no demonstrable left-to-right shunt suggesting that the pulmonic stenosis is of a higher degree and/or the ventricular

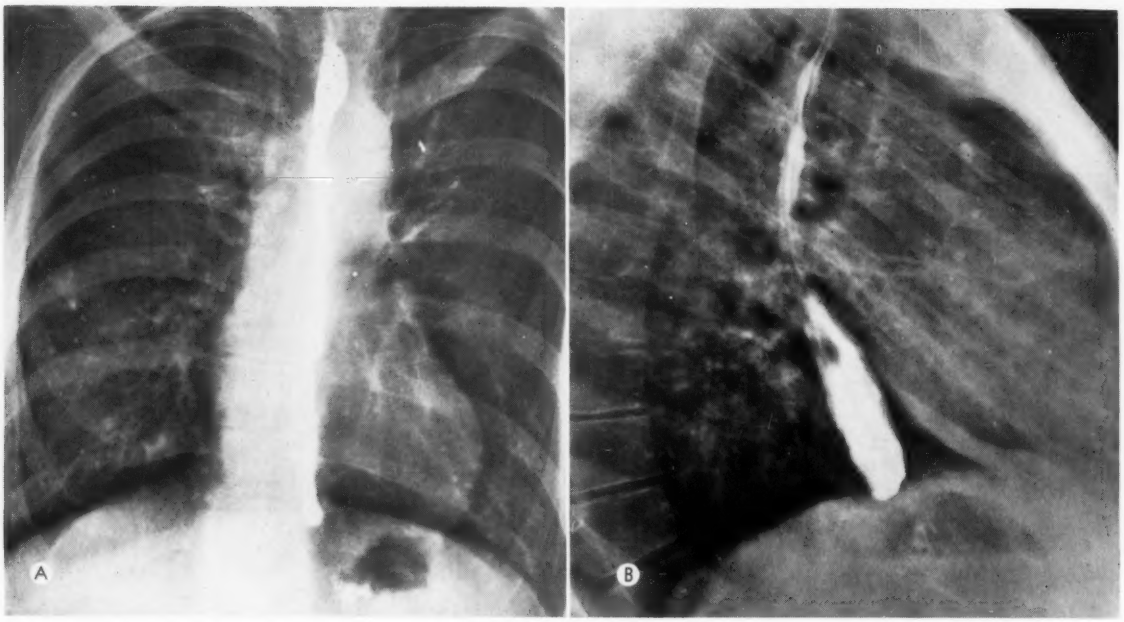


FIG. 5. (*A* and *B*) "Classic" appearance of the tetralogy of Fallot. The heart is small with uptilted apex, concavity in the left mid-border indicating infundibular stenosis. Bronchial collateral circulation is evident.

septal defect is small. In these patients one might justify the use of the descriptive term "balanced shunt" implying that the ventricular septal defect is virtually "closed" to the shunting of significant volumes of blood because of the "balance" established with the obstruction at the pulmonic valve. The roentgen findings in these 2 cases similarly coincide with the physiologic data. In patients of this type there is no evidence of left-to-right shunting and the pulmonary vasculature may be normal or diminished. It is in these cases where one is more apt to suspect the presence of pulmonic stenosis.

The variability of the clinical and roentgen findings in tetralogy of Fallot demonstrates the necessity of accurate evaluation of not only the structural defects but also their influence upon cardiac physiology. It is only with such information that an intelligent surgical approach to the problem can be made.

## SUMMARY

The tetralogy of Fallot can no longer be considered a congenital cardiac defect with uniform clinical and roentgen manifestations. To illustrate this point, 4 patients with acyanotic tetralogy of Fallot are presented with the appropriate catheterization and roentgen findings. Not only are the clinical features extremely variable but the roentgen findings are in accordance with the predominant defect; namely, the pulmonic stenosis or the ventricular septal defect. Tetralogy of Fallot must now be considered in the differential diagnosis of acyanotic congenital heart disease.

Theodore E. Keats, M.D.
University of Missouri
School of Medicine
Department of Radiology
Columbia, Missouri

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Vol. 82, No. 3

# THE DYNAPULSE METHOD OF ULTRASHORT (MILLISECOND) TIMING AND IMPROVED RAPID IMPULSE TIMING IN MEDICAL ROENTGENOGRAPHY

*By* ROBERT B. FUNCH, M.D.*
PHILADELPHIA, PENNSYLVANIA

WITH the use of impulse timing and modern heavy-duty rotating anode tubes, roentgen-ray exposures as short as 1/60 (0.017) second have long been practical and have proved adequate for routine medical roentgenography even where involuntary motion is involved. The exposure requirements for a sharp image in angiocardiography, however, are more severe and have led to the development of ultrashort-timing equipment to eliminate or minimize the blurring due to motion of contrast material, the cardiac valves and the heart itself. Such a system of ultrashort timing will be described and its clinical applications will be discussed and compared with an improved impulse timing system.

## IMPULSE TIMING

Timing in a conventional high-powered roentgen-ray generator is accomplished by mechanical switching in the primary circuit of the high tension transformer phased with the sine wave of the power supply to avoid harmful voltage and current transients (Fig. 1). Under these circumstances, exposure intervals are generally limited to integral multiples of a one cycle period of the alternating current power supply. With the usually available 60 cycle power supply in this country, the most rapid exposure is 1/60 (0.017) second. Some recently developed systems permit the use of an odd number of half cycles with minimal exposure times of 1/20 (0.05) second.

The use of half cycle exposure of 1/120 (0.008) second is possible, but presumably represents the limit with conventional circuitry without raising the frequency of the power-line supply. An impulse timer modified to permit such half cycle exposures has been used at the Germantown Hospital since 1955.* The photographically effective portion of such exposures is of the order of 1/240 (0.004) second because of the sine wave shape of the voltage pulse and is referred to in the remainder of this presentation as "rapid impulse" exposure.

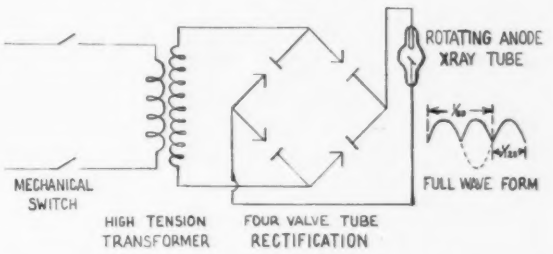


FIG. 1. Schematic diagram of conventional roentgen-ray circuit with full wave rectification. Note the location of the timer in the primary circuit of the high tension transformer and the voltage wave form at the roentgen-ray tube.

Departing from conventional one phase circuitry, Kjellberg *et al.*,[4] Rudhe,[6] and others have obtained excellent angiocardiograms using a three phase generator with a thyratron contactor to obtain exposures as rapid as 1/333 (0.003) second.

## ELECTRONIC SWITCHING TUBE TIMING (DYNAPULSE)

Development of an alternative approach to even shorter exposures was stimulated by Dotter[1] and others who felt that exposures of one to three milliseconds would further improve the diagnostic yield in angiocardi-

* Engineered by the Franklin X-Ray Corporation, Philadelphia, Pennsylvania.

* Chief of Radiology, Germantown Dispensary and Hospital; Associate in Radiology, Temple University School of Medicine.

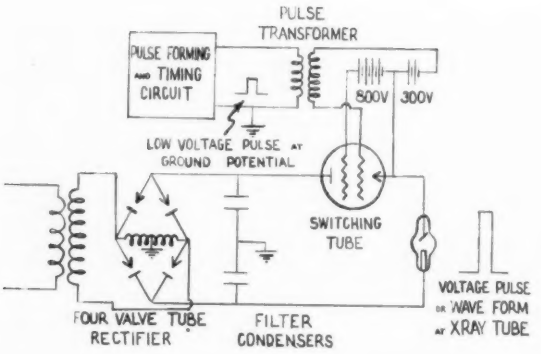


FIG. 2. Schematic diagram of circuit of electronic switching tube system (Dynapulse). Note the timer in the secondary circuit of the high tension transformer, the condensers and the "square" shape of the voltage pulse at roentgen-ray tube (after Rogers[5]).

ography. In the Dynapulse timer* making and breaking the circuit occurs in the secondary circuit of the high tension transformer using as a contactor a controlled electron-flow vacuum tube rather than any form of mechanical switch (Fig. 2).

The switching tube is a tetrode and is used with a conventionally rectified roentgen-ray generator having a filter condenser across its output. Prior to exposure, the condenser is charged to its full open circuit voltage and this power is released to the roentgen-ray tube by the switching tube eliminating the need of synchronization of the exposure with the sine-wave power supply. The entire electron switching system including the condensers is mounted as a unit between the pre-existing conventional generator and the roentgen-ray tube and can be cut in or out of the circuit at will by remote control. Exposures of 1/2,000 (0.0005) second to 5 milliseconds are possible with such a unit used at Germantown Hospital since 1955, though the shortest exposures advocated by the manufacturer are 1/1,000 (0.001) second. The latter are four times faster than the shortest possible exposure with impulse timing. Switching tube system exposures are referred to in the remainder of this presentation as millisecond or Dynapulse exposures.

Our Dynapulse timer is tied in with an exposure-programming device and rapid serial film changer† so that only a single button need be pushed for a programmed sequence of millisecond exposures under control of the angiographic device. A recycle circuit signals the film changer at the instant the exposure is completed to cause it to perform its film changing function and initiate the succeeding steps in the exposure sequence control.

The voltage pulse applied to the roentgen-ray tube as illustrated (Fig. 2) does not exhibit a sine-wave form, but begins and terminates abruptly with a drop during the pulse of 4 kv. per millisecond. For this reason, photographically effective and useful radiation is produced throughout the full interval of exposure.

It is emphasized that a direct comparison of milliampere-seconds between impulse timing and Dynapulse timing is not possible. This is because of the differences in usable radiation during the interval of exposure and because milliammeter readings with conventional timing represent effective values of the pulsating current while in Dynapulse timing the current is constant and not pulsatile.

The electronic switching system has been used with a 2 mm. focal spot rotating anode tube with 85 kv. peak at 1,000 ma. and 1 millisecond or 115 kv. peak at 600 ma. and one millisecond. Proposed circuitry improvements and the use of a 2.5 mm. focal spot rotating anode tube will increase permissible exposures by 90 per cent.

## EXPERIMENTAL APPLICATION AND COMPARISON

To critically compare the unsharpness of image due to motion with the two timing methods, roentgenography of a moving mechanical test object was performed. A fine wire mesh driven on a spinning disk by synchronous motors of known speed was

* Developed by the Machlett Laboratories, Incorporated, Springdale, Connecticut.

† Developed by the Franklin X-Ray Corporation, Philadelphia, Pennsylvania.

used for this purpose (Fig. 3). It should be noted that this is a very severe test, since the fine wire mesh presents greater detail than one would expect to encounter in the analysis of lung roentgenograms or angiocardiograms and is more comparable to fine trabecular detail of bone. Unsharpness of the image at the rapidly moving periphery of the revolving screen and "stoppage" of motion near the hub of the axle are apparent. The speed of the circular path where unsharpness of the mesh is grossly eliminated is readily determined by multiplying the known speed of the disk by the circumference of the circle.

$$\text{Speed (cm./sec.)} = \frac{\text{RPM}}{60}\ \text{disk} \times 2\pi r\ \text{(cm.)}.$$

Using this method, motion of about 80 cm. per second is "stopped" by 1/1,000 second exposure and of 20 cm. per second by 1/120 (0.004 effective) second exposure with "rapid impulse timing." This is a ratio of 4:1 as expected since the former exposure is four times faster. If a less critical end point such as the sharpness of the edge of the screen is used, then speeds up to 120 cm. per second are "stopped" by Dynapulse timing. Under physiologic conditions, blood flow in man is thought to approach 100 cm. per second, comparable to speeds "frozen" by a 1/1,000 second exposure, but the rate of flow past stenotic valves probably attains speeds of up to 600 or 700 cm. per second.[6]

## CLINICAL ROENTGENOGRAPHIC APPLICATION AND COMPARISON

A. *Plain Roentgenography*. The switching tube system has functioned reliably in clinical roentgenographic application. Where motion is not encountered (spine, extremity, etc.), there is no difference in the sharpness of the roentgenograms exposed with Dynapulse, "rapid" impulse or conventional impulse techniques other than that due to the smaller focal spot size possible with the last mentioned.

Radiation exposure, however, is reduced

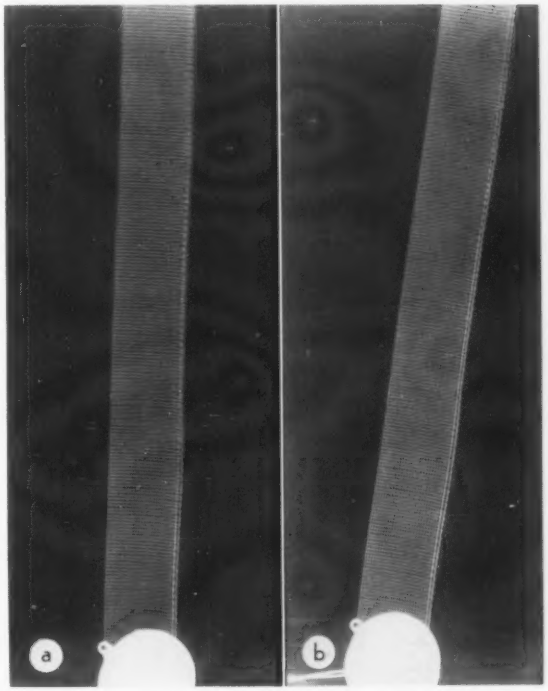


Fig. 3. Roentgenograms of the mechanical test object moving at known speeds exposed (*a*) at 1/120 second and (*b*) at 1/1,000 second.

with the "harder" beam of the ultrashort exposures because of their square voltage wave form of relatively constant potential. Kirka[3] has found it necessary to use approximately only one third the output with 1 millisecond switching tube exposures as with 1/60 (0.017) second impulse exposures to produce roentgenograms of comparable density. Further reduction in exposure can be expected with millisecond timing since fewer retakes should be necessary for blurring from patient movement, especially in pediatric roentgenography.

The reduction in health hazard from radiation exposure was not planned for primarily as such, but has been a side development of the Dynapulse method. It is, nevertheless, a very significant and important contribution, especially in view of the increasing efforts to confine medical radiation exposure to the necessary minimum.

A comparison of chest roentgenograms exposed at 1/1,000 second, 1/120 second and 1/60 second was carried out to evalu-

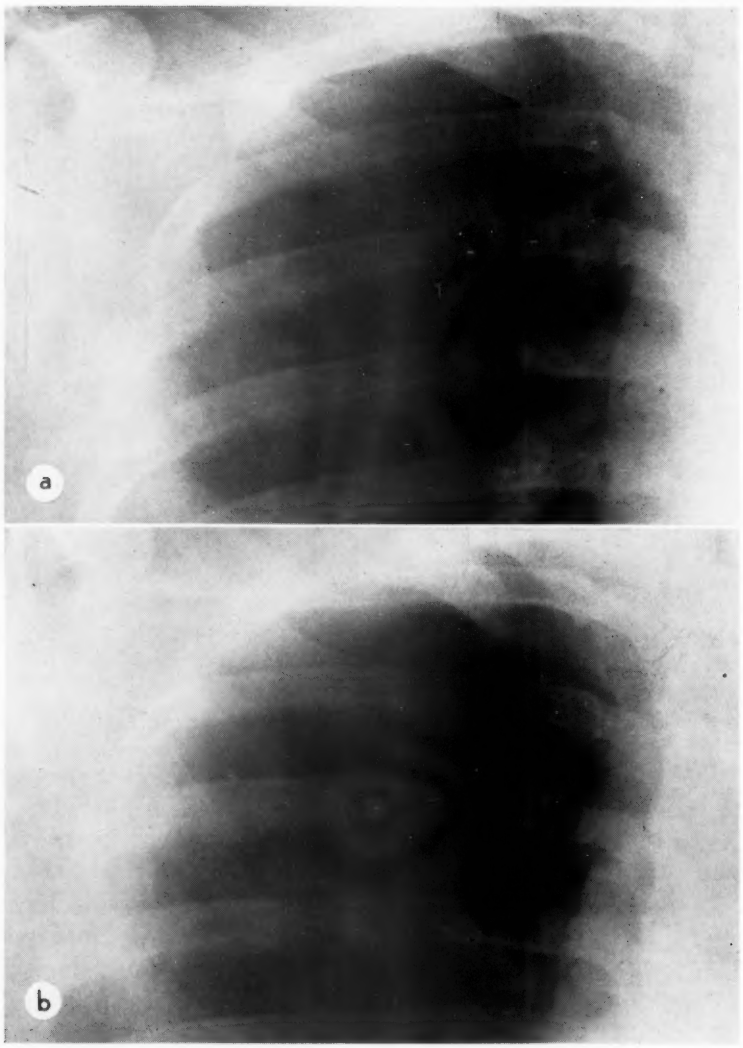


FIG. 4. Roentgenogram of adult female chest exposed (*a*) at 1,000 ma. and 1/1,000 second and (*b*) at 50 ma. and 1/60 second, using 75 kv. peak. No difference in detail of pulmonary markings is noted.

ate the comparative ability of the ultrashort, "rapid" impulse and conventional impulse timed exposures to record the sharpness of the pulmonary vascular tree (Fig. 4). The group studied ranged from age three months to adulthood and included normal and diseased lungs. Four radiologists interpreted the films independently without knowledge of the identity of the Dynapulse films and were unable to detect any difference in the amount or detail of lung markings recorded with the several exposure times. Therefore, conventional timing seems as valuable as the Dynapulse and "rapid" impulse timing in "stopping" motion due to pulsation of pulmonary vessels.

B. *Angiocardiography.* To compare the relative effectiveness of Dynapulse and "rapid" impulse timed exposures in "stopping" motion in angiocardiography, experimental animals were used since multiple control examinations of the same child or adult did not seem justified as they would require otherwise unnecessary repeated injections, radiation exposure and patient risk. Therefore, successive contrast examinations were carried out on dogs at

1/1,000 second and also at 1/120 second. They could be critically compared since all other factors remained unchanged including film centering, patient size, pulse rate, anesthetic, concentration and volume of contrast medium, injection time and film processing. In the normal animals examined, the 1/1,000 second timed and 1/120 second timed venous angiocardiograms were of equally excellent quality (Fig. 5). There was no objective discernible difference in their sharpness. The opened and closed pulmonary and aortic valves, the mitral valve, the muscular trabeculae of the heart, the coronary arteries and the great vessels were clearly demonstrated. When left ventricular injections of contrast were used, the influx and mixing of the nonopacified left atrial blood with the bolus of contrast material in the ventricle was visualized equally well with the two methods (Fig. 6). This phenomenon of mixing of opacified and nonopacified blood in the left ventricle can be considered a physiologic "jet" comparable in many ways to the jet or shunt of blood through an intracardiac defect. For this reason, it is probable that Dynapulse and "rapid" impulse exposures would be equally effective in the visualization of congenital or acquired shunts, though animals with experimentally produced intracardiac defects have not actually been examined as yet with the comparative techniques.

A superiority of Dynapulse timing over "rapid" impulse timing was demonstrated, however, when particulate contrast material (iodochloral) was used. Unsharpness due to motion of oil droplets in the proximal aorta at the moment of maximal propulsion seen on 1/120 second films was eliminated on 1/1,000 second films. Since, at present, there are no safe particulate contrast agents, this advantage appears to have little clinical value. It should, however, be useful in the physiologic investigations of the velocity, turbulence, etc. of blood flow in the experimental animal, because the path of an individual droplet could be followed on rapid serial films exposed and studied at known intervals.

Clinical application of the switching tube and the "rapid" impulse timing systems at Germantown Hospital in patients with congenital heart disease has not been extensive. As indicated before, a repeat angiocardiogram in the same projection with the alternative timing method was not done on the same patient because of the added risks and, therefore, a direct comparison in the same patient was not possible. On occasion, however, a patient had an angiocardiogram in the anteroposterior projection exposed at 1/1,000 second and in the lateral projection at 1/120 second (Fig. 7). Interpretations of these human studies to date have not revealed any obvious advantage of the greater motion "freezing" ability of the millisecond exposure over 1/120 second exposures in regard to sharpness of contrast or the ability to demonstrate intracardiac anatomy or pathologic defects. Lesions studied included tetralogy of Fallot, pulmonary stenosis, atrial septal defect, abnormal connection of pulmonary vein and great vessel anomalies (Fig. 8). It is possible that a more extensive clinical trial might reveal an increased yield in the diagnosis of shunts, defects or valve abnormalities with Dynapulse exposures, but present experience suggests that impulse timed exposures of 1/120 second meet the practical speed requirements for these purposes in direct roentgen angiocardiography. Both exposure techniques are undoubtedly superior to slower conventional timing methods. Despite the statement of Dotter to the contrary, it is felt that sharp detail with ultrashort exposures does not obviate the need for rapid serialographic studies.

Experience with aortography and cerebral angiography indicates no diagnostic advantages of millisecond timing over conventional methods and the use of the Dynapulse unit depends on its availability and the desire to reduce radiation exposure to the patient and medical personnel.

The use of the ultrashort timing in inefluorography with image amplification

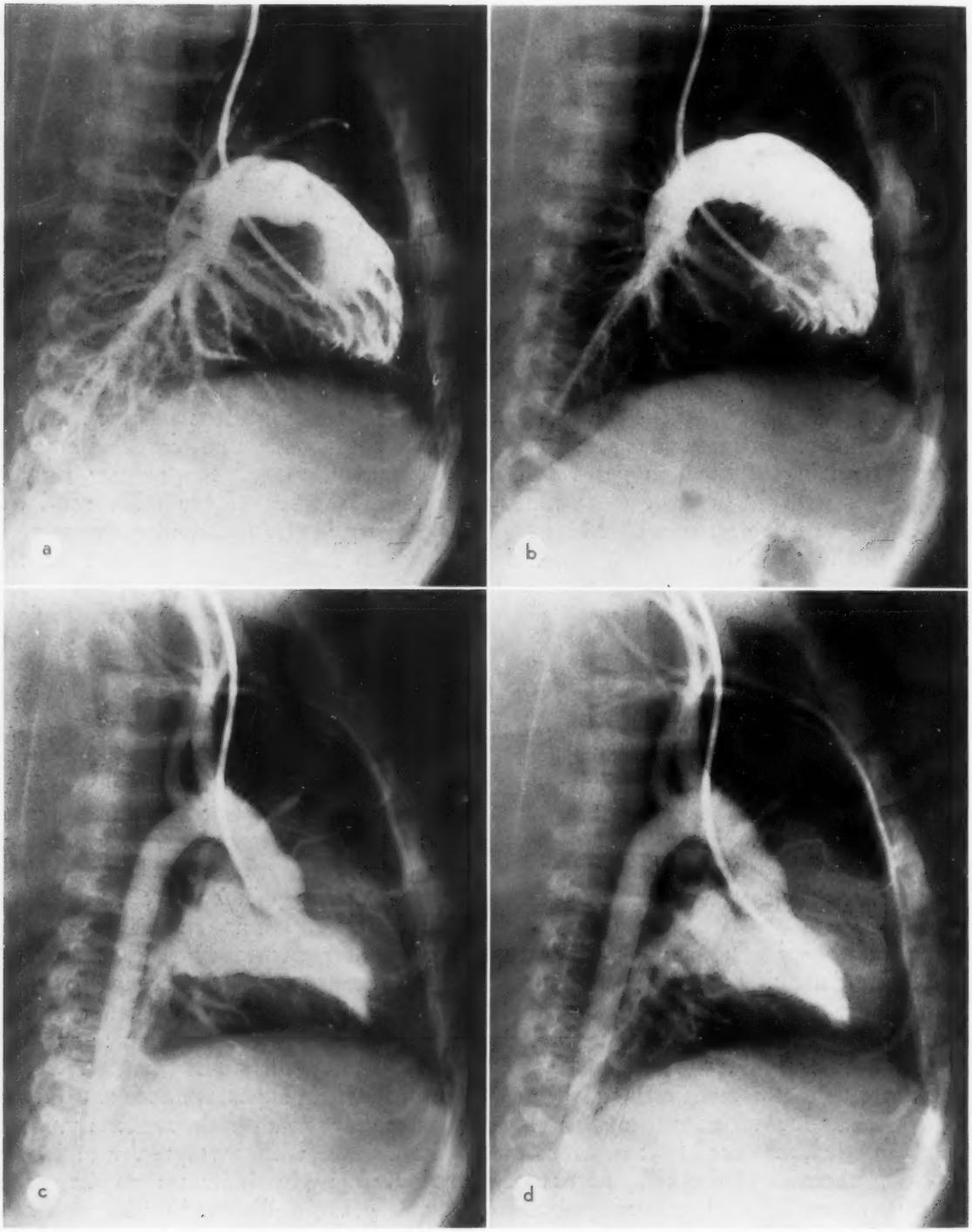


FIG. 5. Dextro-angiocardiograms of normal dog exposed (*a*) at 1/120 second and (*b*) at 1/1,000 second. Levo-angiocardiograms of same animal exposed (*c*) at 1/120 second and (*d*) at 1/1,000 second. The contrast medium was injected into the right ventricle. Note the equally excellent detail of the opaque bolus, muscular trabeculae of the heart, cardiac valves, coronary arteries and the great vessels with each method.

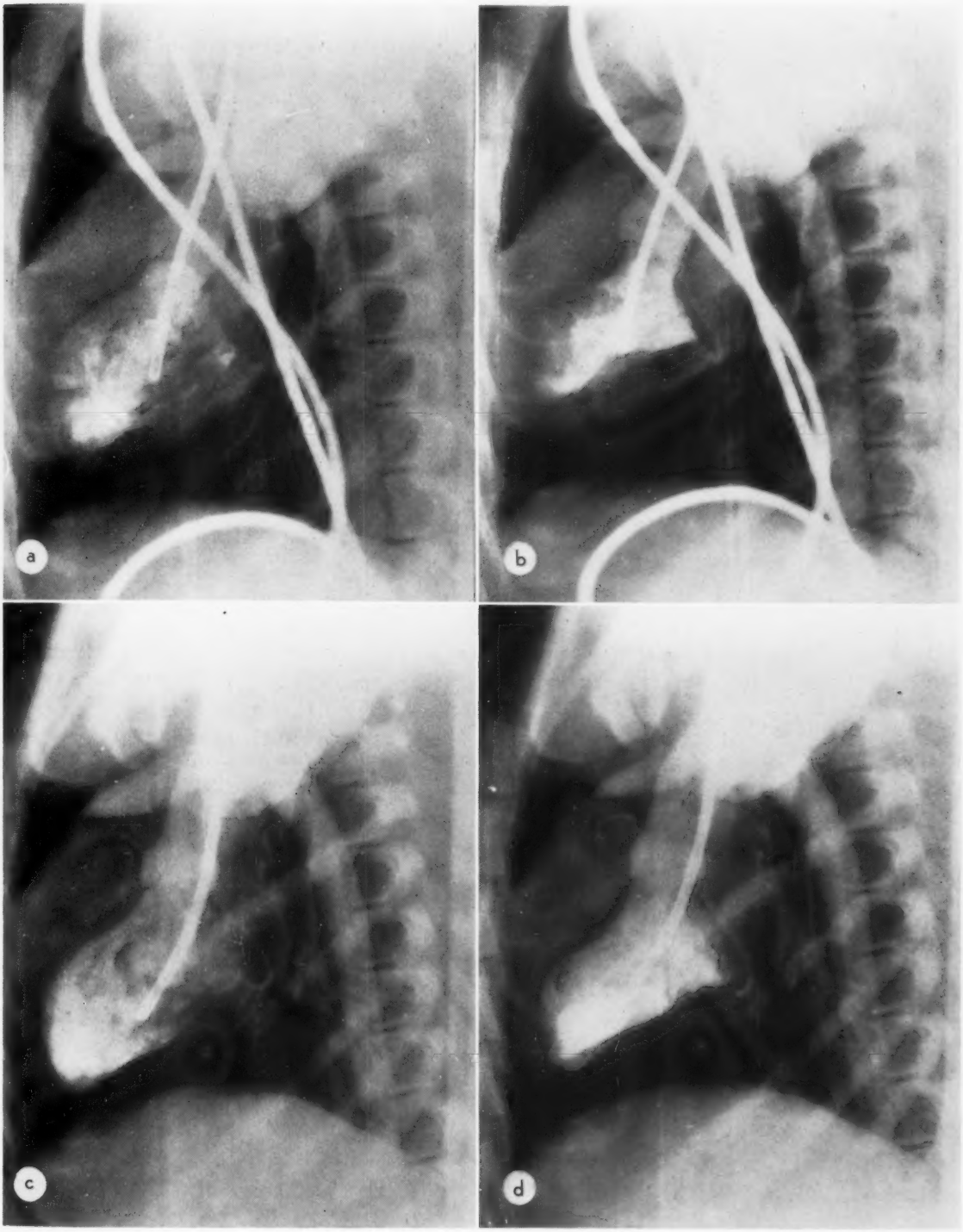


Fig. 6. Levo-angiocardiograms of normal dog exposed (*a* and *b*) at 1/120 second and (*c* and *d*) at 1/1,000 second, with the contrast medium injected into the left ventricle. Note the mixing of physiologic jets of nonopacified blood from the atrium simulating abnormal shunt.

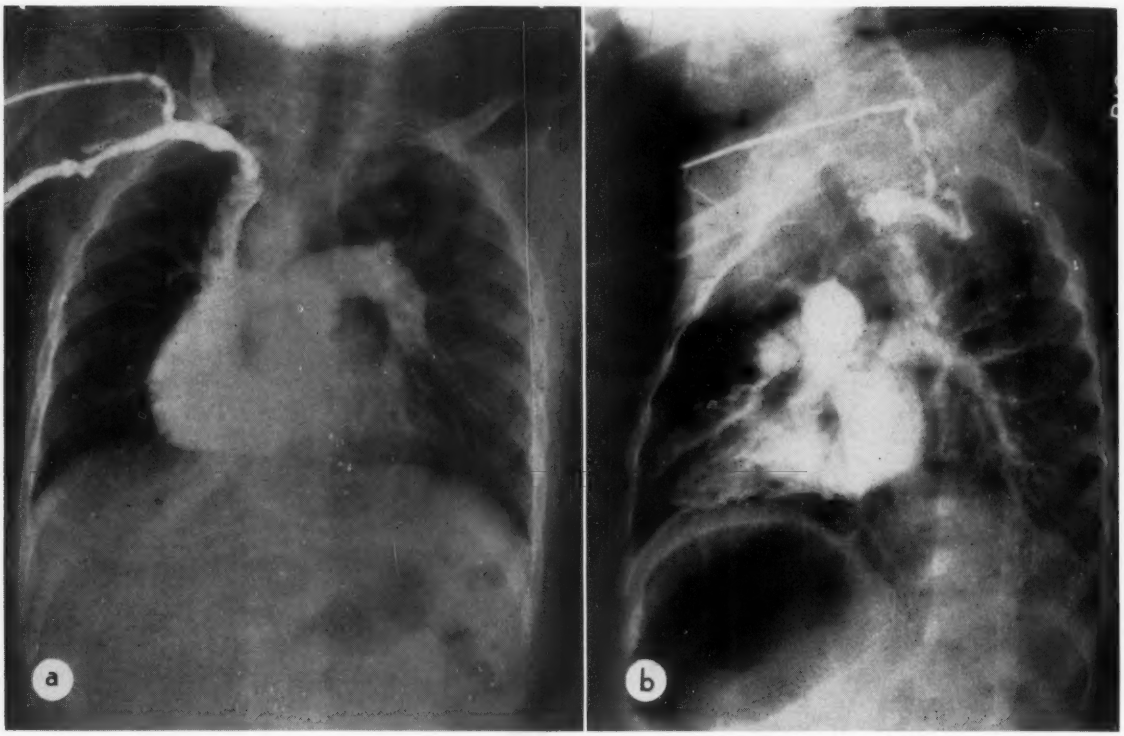


FIG. 7. Angiocardiograms in a two year old female with tetralogy of Fallot exposed in anteroposterior projection (*a*) at 1/1,000 second and in lateral projection (*b*) at 1/120 second. Note the excellent detail of the right ventricular outflow tract with stenosed ostium infundibuli, "third" ventricle and pulmonary valvular stenosis clearly identified at 1/120 second exposure. A third site of obstruction in the pulmonary artery is revealed on the underexposed anteroposterior film at 1/1,000 second.

may prove to be a valuable application of the switching tube system. The Germantown unit is capable of only 12 exposures a second and is therefore not suited for cine techniques. The manufacturer, however, has modified the system to permit up to 60 exposures a second synchronized as the slave to the control of the movie camera shutter. Reduction in radiation exposure with this method is obvious since radiation is eliminated during the period the camera shutter is closed and because of the more homogeneous nature of the Dynapulse roentgen-ray beam described before.

## CONCLUSIONS

1. The exposure requirements for a sharp image in angiocardiography necessitate faster timing than in routine medical roentgenography to minimize blurring due to motion. An electronic switching tube method of ultrashort 1/1,000 second exposures has been reviewed and compared with an improved 1/120 second impulse timing system in this respect.

2. The Dynapulse tetrode tube method performed reliably in experimental and clinical application and is four times faster than the photographically effective portion of 1/120 second impulse timed exposures.

3. Dynapulse timing revealed no diagnostic superiority, however, in routine roentgenographic examinations including chest roentgenography, in cerebral angiography or in angiocardiography in normal dogs.

4. Both timing methods are superior to slower conventional methods for angiocardiography in congenital heart disease and to date exposures of 1/120 second have

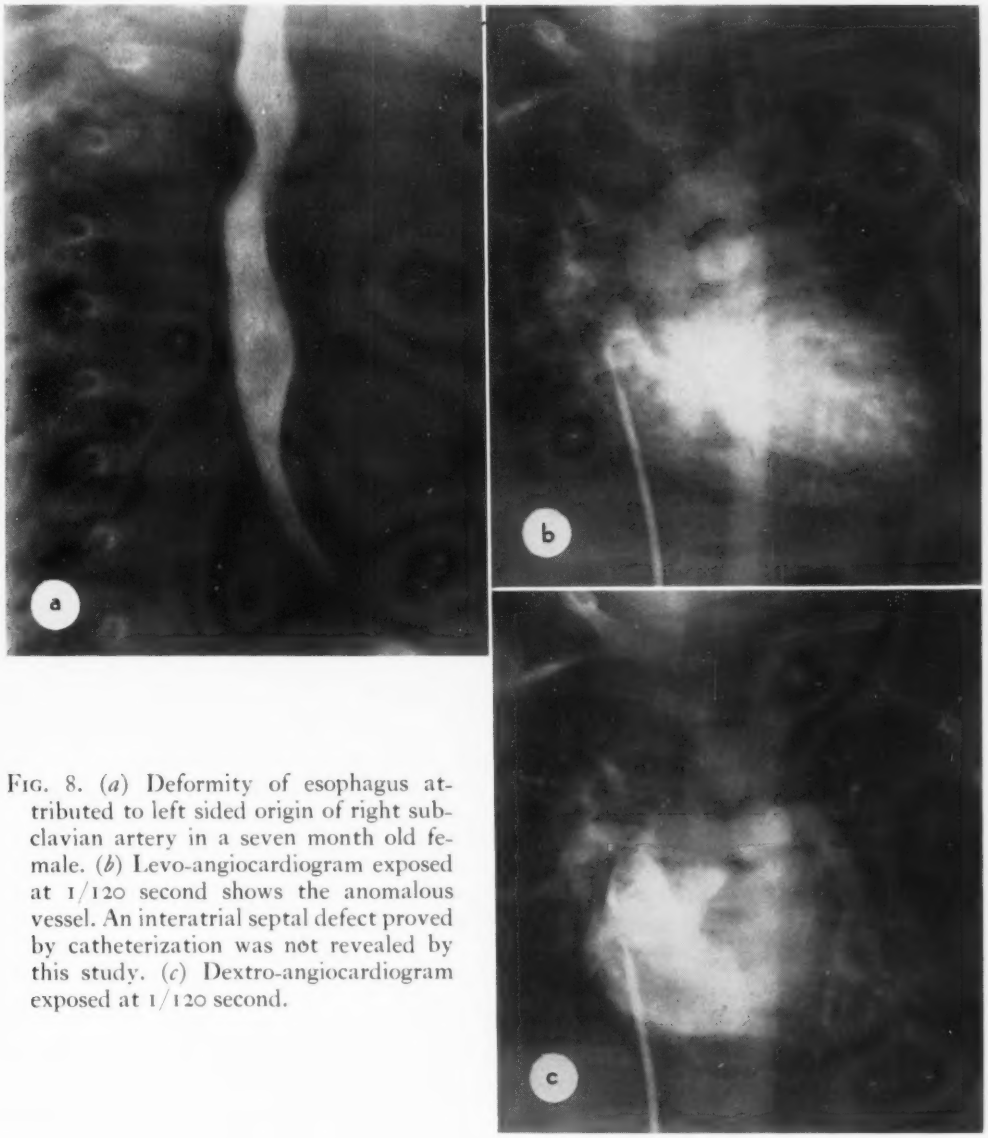


Fig. 8. (*a*) Deformity of esophagus attributed to left sided origin of right subclavian artery in a seven month old female. (*b*) Levo-angiocardiogram exposed at 1/120 second shows the anomalous vessel. An interatrial septal defect proved by catheterization was not revealed by this study. (*c*) Dextro-angiocardiogram exposed at 1/120 second.

seemed to meet the practical speed requirements for direct roentgenography as well as do 1/1,000 second exposures.

5. Superiority of 1/1,000 second exposures has been found with particulate contrast material, but for the present this must be restricted to the physiologic study of hemodynamics in experimental animals.

6. An important advantage of the switching tube system is reduction in health hazard due to radiation exposure.

7. Dynapulse timing is particularly adaptable for use in cinefluorography at camera speeds of up to 60 frames per second.

8. Ultrashort exposures do not obviate the need for rapid serialography in angiocardiography.

Robert B. Funch, M.D.
Germantown Hospital
Philadelphia 44, Pennsylvania

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# ROENTGENOLOGIC CONSIDERATIONS OF CAPILLARY BRONCHIOLITIS

*By* DONALD A. KOCH, M.D.
PORT HURON, MICHIGAN

CAPILLARY bronchiolitis has been frequently diagnosed by clinicians for many years. It has also been recognized for its roentgen features by many radiologists. However, there has been little written in the radiologic literature and little has been done with this syndrome in scientific exhibits. Thus, the primary purpose of this paper is to bring it into proper focus roentgenologically and to point out certain typical features which produce detectable abnormality on the roentgenogram of the chest.

Capillary bronchiolitis can be described as an acute clinical syndrome characterized by respiratory distress and emphysema in an acute respiratory infection in infants. The fact that it is an acute condition and that it is found in infants distinguishes it from lung changes in cystic fibrosis of the pancreas and from emphysema due to ordinary allergy. Similarly, the lung changes can be readily differentiated from overinflation of the chest due to exposure of the film during maximum inspiration.

Although it is generally accepted that the roentgenologic description of one case practically never fits closely the description of another case, there are certain features which are common to most. When they are present in an acutely ill infant, bronchiolitis must be placed at the top of the list in the differential diagnosis. A review of the clinical pattern of the syndrome will help to better understand the roentgen changes observed.

## ETIOLOGY

There is no known specific causative agent or organism. General opinion favors the view that the syndrome is produced by any organism invading the respiratory tract in infants. Various organisms have been suspected as the etiologic agents, among them the *Hemophilus influenzae* and the virus producing distemper in puppies, but none has as yet been isolated.

## INCIDENCE

The disease occurs most commonly during the first six months of life and is rare after the age of two. Approximately 75 per cent of the cases are observed during the first year of life. Stiller[9] found 95 per cent of the cases in the first six months of life. The incidence is highest in the winter and spring months; it changes sporadically, but is usually increased during epidemics of upper respiratory infections in older children and adults. The condition is noted with equal frequency in both sexes and there are no known racial differences.

## CLINICAL FEATURES

The usual history discloses that the infant has been ill with an upper respiratory infection for a few days to a week. At this time, nasal discharge, cough, sneezing and mild constitutional symptoms are present. An abrupt change occurs during the onset of pulmonary involvement. The infant becomes violently ill with severe respiratory distress and signs of obstructive emphysema. Both respiration and pulse rate become accelerated, the breathing is labored and cyanosis may appear. There is usually a dry cough and signs of dehydration may follow. The child is apprehensive and restless. The temperature is generally less than 100° F. but may rise to 101° F. or 102° F.

## PHYSICAL FINDINGS

Examination reveals a slightly cyanotic, desperately ill infant. The chest is overexpanded and there is obvious overactivity of the muscles of respiration. The chest is hyperresonant and the expiratory sounds are prolonged. There are diffuse rales and rhonchi on auscultation. The air exchange is limited and the breath sounds are di-

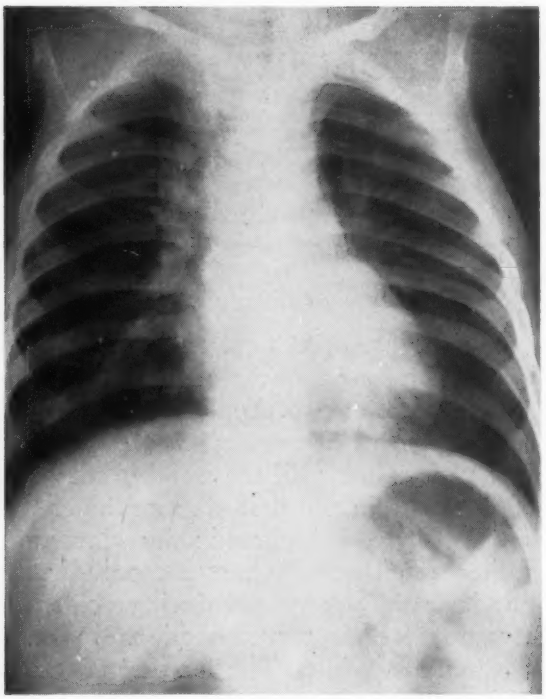

FIG. 1. "Too normal" appearing chest roentgenogram. This infant expired less than twenty-four hours after this roentgenogram was taken. Autopsy showed that death was due to bronchiolitis.

minished. The pulse and heart rate are rapid. The child is often dehydrated. A survey of nose and throat shows inflammatory changes with some congestion. The liver and spleen may become palpable because of displacement by the depressed diaphragms.

### LABORATORY DATA

Cultures of sputum are not specific and blood studies are of limited value. Most observers believe that there is an early leukocytosis followed by normal or depressed white blood cell count. The differential blood cell count is not unusual. Mild to moderate acidosis may be present and the oxygen saturation of the arterial blood is reduced according to High.[4]

### PATHOLOGY

In spite of the fact that the prognosis generally is very favorable and the pathologic changes are few, certain features appear to be characteristic. Grossly, the lungs are strikingly emphysematous. There are scattered areas of atelectasis and consolidation. The air passages from the pharynx to the bronchi show hyperemia. No obstructions are noted in the trachea or major bronchi. Microscopic examination reveals a severe thickening of the walls of the small bronchi and bronchioles as a result of lymphocytic infiltration. Mild inflammatory exudate extends into the adjoining alveolar septa and moderate infiltrates of neutrophils are found in bronchiolar lumina and alveolar spaces. There is severe emphysema present in addition to the patchy areas of atelectasis and bronchopneumonia.

### ROENTGEN CHANGES

Often the radiologist is confronted with a frontal roentgenogram of the chest in which the lung fields appear unusually clear and the diaphragms are pulled down at the height of inspiration. The impression is one of optimal technical detail and exposure at the most perfect time in the respiratory cycle. Paradoxically, this so-called "too normal" appearing chest roentgenogram is found to be the roentgenogram of an acutely and desperately ill infant. Then, as the infant's clinical course improves, symptoms subside, the apparent changes and the abnormal features on the roentgenogram taken at the height of the illness become evident in retrospect. This is shown to good advantage in Figure 1. The relative normal appearance of this frontal roentgenogram of the chest belies the fact that this infant expired less than twenty-four hours later with bronchiolitis. Figure 2, a photomicrograph of the autopsy specimen of this patient, shows the pathologic changes in a bronchiole and the adjacent lung.

We have found that roentgenograms in inspiration and expiration in both frontal and lateral views give the most information. For this reason, we have adopted this procedure as routine. Obtaining roentgenograms in expiration is obviously difficult if not almost impossible in these cases, whereas in most other chest conditions it seems difficult to obtain roentgenograms of

infants' chests in inspiration. Nevertheless, even slight differences in the depth of inspiration in bronchiolitis bring out inequalities in aeration which are characteristic of this disease. Furthermore, they bring out the pneumonic and atelectatic densities to better advantage and show the influence of the tightly distended lung on the cardiac silhouette.

On the frontal roentgenogram in inspiration the lungs show increased radiolucency with depressed diaphragms where the variations in the crurae produce a scalloped effect (Fig. 3). The costophrenic sulci are broadened and the acuity of the angles is obliterated. The vascular shadows appear to be stretched out, particularly from the hili to the median bases in the overdistended lung. On the frontal roentgenogram in expiration, the obstructed portions of the lungs become more apparent as the unobstructed parts lose their radiolucency. Atelectatic areas and segments of consolidation become more obvious.

Finally, a very interesting change is often noted in the cardiac shadow. The overdistended lung apparently exerts enough pressure on the heart to reduce its size, whereas in the normal chest the heart shadow increases in size during expiration. This paradoxical change in heart size is probably a sign of an added burden on the already overworked heart.

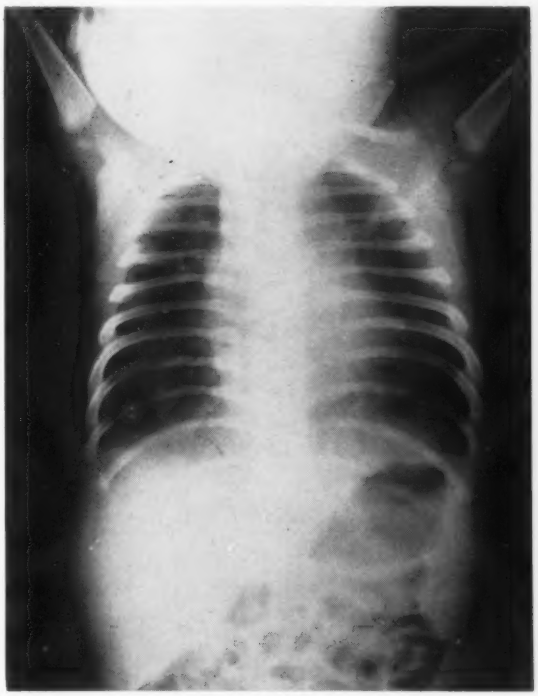

Fig. 3. Frontal roentgenogram. Increased radiolucency of the lung fields. The stretched vascular shadows and the severely depressed diaphragms with scalloped effect are well demonstrated.

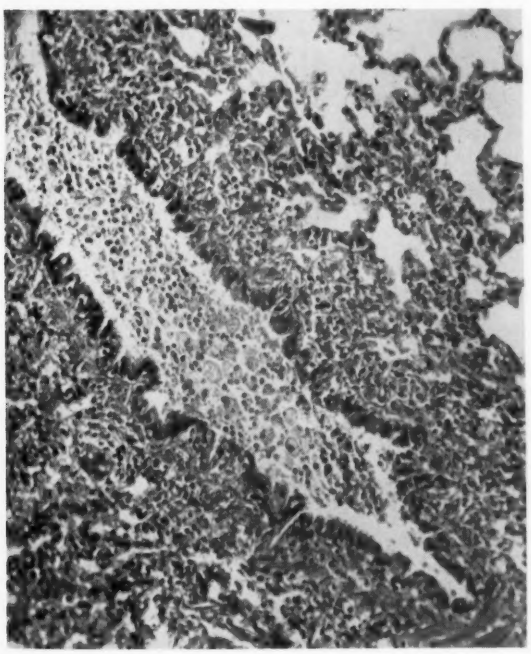

Fig. 2. Photomicrograph of a section of the lung of the case shown in Figure 1. Abundant inflammatory reaction in lumen and walls of bronchioles. Focal areas of atelectasis in adjacent lung parenchyma. (×190.)

On the lateral roentgenogram of the chest, perhaps the earliest evidence of lung abnormality is the depressed surface of the diaphragm in the posterior half of the chest. This is often a reversal of the normal convexity (Fig. 4). Its recognition is easy when looked for and almost invariably present. Other interesting signs are the increased anteroposterior diameter of the chest and the bulging of the lung anteriorly, especially into the intercostal spaces, in particular if the roentgenogram is slightly oblateral, as described by Benward.[1] There frequently appears to be an increase in the space between the anterior rib ends and sternum (Fig. 5).

Because in infants the upper abdomen is commonly included on the roentgenogram,

Fig. 4. Lateral roentgenogram showing a reversal of normal convexity of the superior surface of the diaphragms, especially posteriorly.

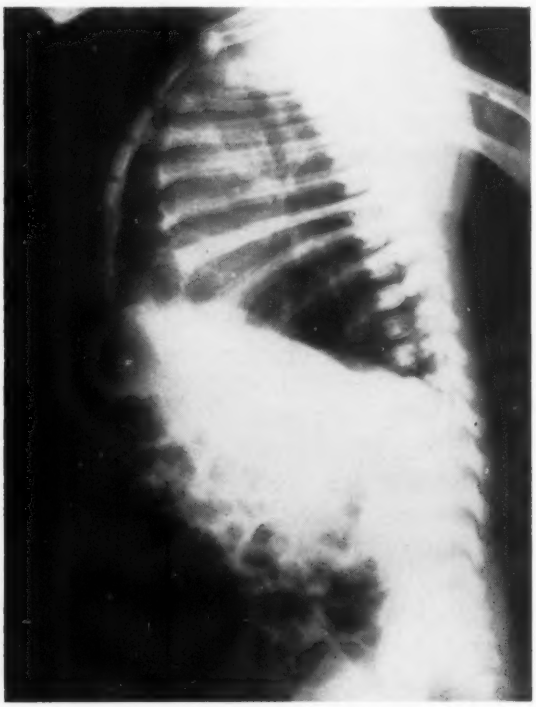

Fig. 5. Lateral roentgenogram, inspiration. Depressed surface of diaphragms posteriorly and increased space between anterior rib ends and sternum.

the shadows of the liver and spleen appear to be displaced inferiorly by the depressed diaphragms. This is borne out by clinical findings as well. The flanks appear to be bulging laterally, probably for the same reason.

## CONCLUSIONS

1. Capillary bronchiolitis is an acute syndrome in infants with certain clinical signs and symptoms.

2. It produces detectable abnormalities on the roentgenogram which aid in establishing the diagnosis.

3. Roentgenograms in inspiration and expiration in both frontal and lateral projections are necessary for convincing evidence of the presence of this disease and the changes observed enable differentiation from normal and other abnormal conditions.

310 E. Water Street
Port Huron, Michigan

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# DIAPHRAGMATIC HERNIA IN INFANCY

*By* J. O. REED, M.D., *and* E. F. LANG, M.D.
DETROIT, MICHIGAN

THE presence of abdominal viscera in the thorax is usually referred to as diaphragmatic hernia, whether the path is through an actual rent in the diaphragm, a naturally occurring aperture, or through a defect due to faulty development. This paper is based on a series of 48 cases of diaphragmatic hernia, all of which were subsequently confirmed either at the time of operation or at autopsy. Inasmuch as the embryology of diaphragmatic development has already been thoroughly detailed in the literature,[1,7,13,14] only a short summary of this will be given.

## EMBRYOLOGY

The ventral division of the diaphragm takes its origin from the septum transversum, which divides the abdominal viscera from the heart. The septum descends from the neck to the level of the twelfth rib where it joins with the thickened mesodermal cells of the dorsal mesentery. Openings referred to as the pleuroperitoneal foramina are present at the posterolateral aspect of each diaphragmatic leaf and these are closed by the pleuroperitoneal membranes which arise from the posterolateral body walls. This usually occurs about the fourth week, with the right foramen closing slightly earlier than the left.

## CLASSIFICATION

Diaphragmatic hernias in infancy, as observed in the roentgen examination, fall readily into three groups. The largest group, containing 31 of the 48 cases in this series, comprises those which are unilateral, involving the posterolateral muscular segment of the diaphragm. The two smaller groups comprise those involving the esophageal hiatus, of which there are 12 in this series, and those involving the anteromedial or tendinous segment of the diaphragm or septum transversum, of which there are 5 cases.

### GROUP I. PLEUROPERITONEAL HERNIAS

These hernias involve the muscular portion of the diaphragm and are due to lack of fusion of the pleuroperitoneal membrane with the septum transversum, the abdominal contents occupying the greater portion of the hemithorax. In this series, the left side was involved in 24 cases and the right in 7 cases. The ratio of males to females is similar to that in cases previously reported. It has long been recognized[5,8,10] that the mortality is high in untreated patients as the result of cardiorespiratory failure in the newborn period (Fig. 1) and from strangulation of the bowel in older children (Fig. 2). In this series there were 5 deaths in 6 patients not operated upon, and only 4 deaths in 25 patients where the hernias were reduced and the diaphragmatic defects repaired.

The roentgen findings are usually characteristic in almost all cases and have been previously described by many authors. They may be listed as follows and are demonstrated in Figure 3, *A* and *B* and Figure 4, *A* and *B*: (1) The diaphragm on the affected side is seen only in part or not at all; (2) there are multiple radiolucencies representing loops of intestine in the thorax; some may contain fluid levels; (3) there is a shift of the heart and upper mediastinum to the opposite side of the chest; there may be associated compression atelectasis; (4) the esophagus is of normal length; (5) there is a relative or absolute absence of the intestinal gas in the abdomen.

In the first few hours of life this last finding may be very important if the viscera in the chest also do not contain gas.

The use of barium sulfate or iodized oil to identify the stomach or intestine is seldom necessary. However, in a few of our cases, the diagnosis was difficult when the plain roentgenograms only were available. Intestine in the thorax may be difficult to

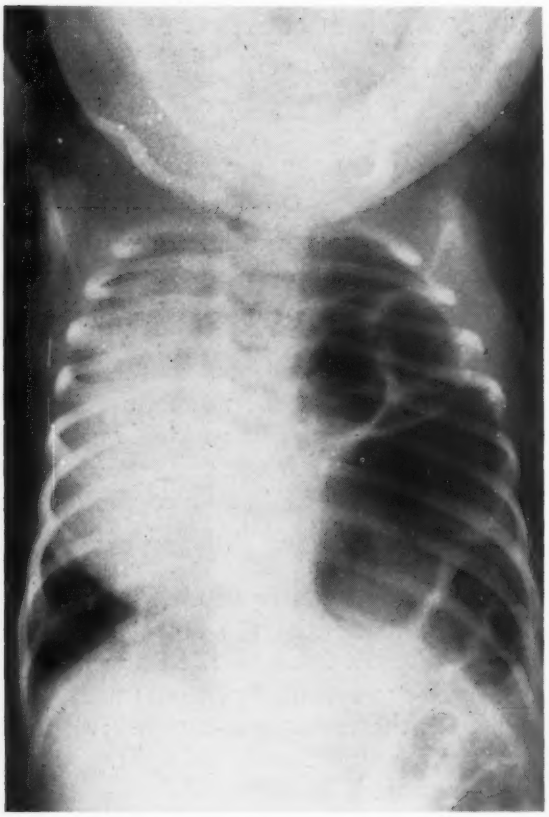

FIG. 1. Large hernia with a sac in a seven day old infant having a defect involving the posterior two-thirds of the diaphragm. Compression atelectasis of both lungs was found at autopsy.

identify if it does not contain gas, as may be seen in Figure 5, *A* and *B*. Pulmonary infections do occur in the presence of diaphragmatic hernia and the pneumonic infiltration may obscure loops of bowel, or it may be misinterpreted as localized emphysema, as in the case demonstrated in Figure 6, *A*, *B*, *C* and *D*. Also the frequently accompanying atelectasis, hypoplasia or aplasia of the ipsilateral lung may be mistaken for pneumonia.

Variation in the apparent size and content of the hernia is to be expected. The upright position during examination of the chest may alter the content of the sac. The surgeon frequently describes solid viscera as herniated when only intestine is demonstrable in the roentgenogram.

Inasmuch as the cardiorespiratory embarrassment may be expected to increase with increasing distention of the stomach and intestine in the presence of large hernias, it is important in the newborn period to delay feeding and to reduce the amount of swallowed air by gastric aspiration until the hernia has been repaired.

It seems probable that a congenital defect may be present in the diaphragm in some cases in which the hernia develops some time after birth. These are of the sliding type, with the herniation present at some times and not present at other times. However, when a large segment of the intestine is involved in the hernia, it is evident that the normal mesenteric attachments have not occurred in the abdomen and that the hernia must date from early fetal life. The role of trauma in the pro-

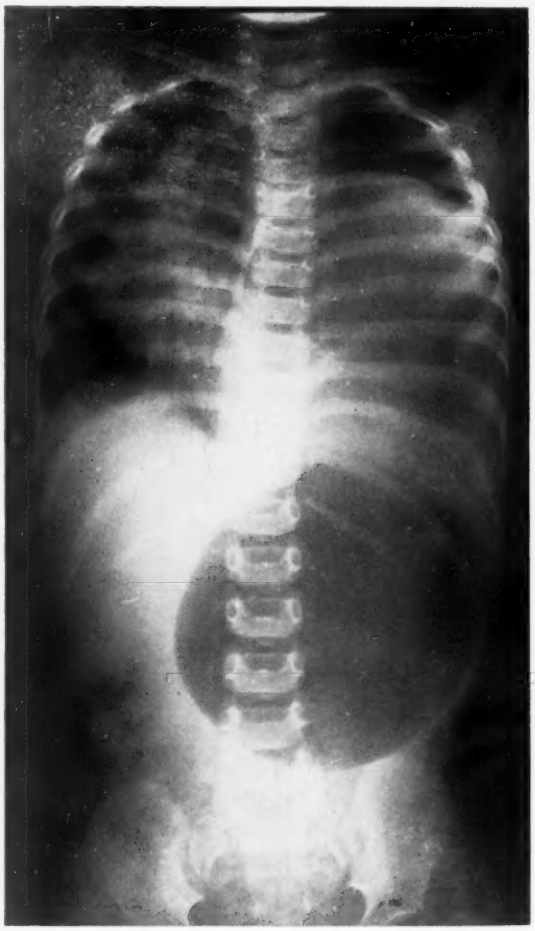

FIG. 2. Volvulus of the stomach associated with a large diaphragmatic defect and hernial sac in an eighteen month old infant. Confirmed at autopsy.

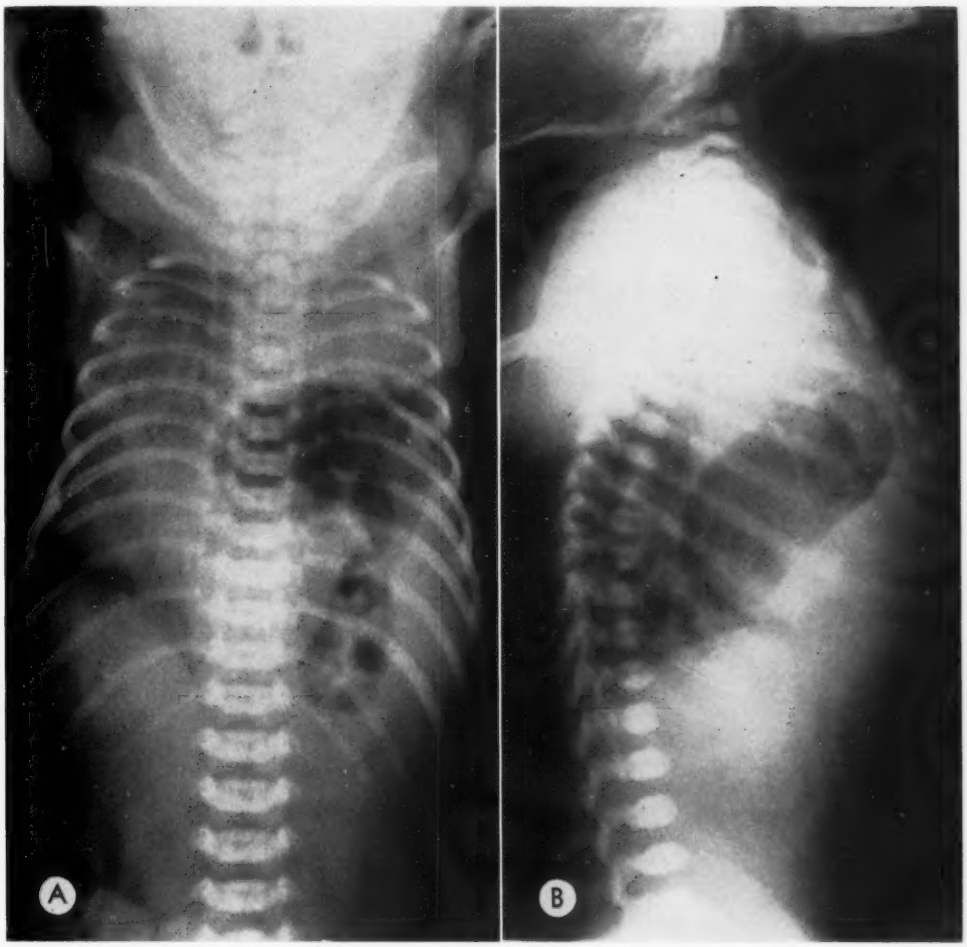


Fig. 3. (*A* and *B*) Left sided hernia seen in a day old infant having an extremely large defect involving almost the entire left diaphragm. There was associated aplasia of the left lung. Death occurred three days postoperatively.

duction of diaphragmatic hernia has not been a problem in this series with the possible exception of one case where the hernia was discovered following a traumatic episode. However, at operation it was the opinion of the surgeon that the defect in the diaphragm, if not the hernia, was congenital in origin.

If the hernias in this group are very large and associated with severe defects in the lung, either developmental or acquired because of the thoracic mass, they are frequently inconsistent with sustained life. These infants do not survive long enough for operative repair and are not seen in a children's hospital. A certain number can be kept alive for a short time, and these can frequently be saved by immediate operation if the pulmonary defect is not too severe or is correctable. Occasionally, a poor prognosis can be made from the initial roentgen examination (Fig. 7, *A* and *B*).

One limitation of the roentgenologic examination is the failure to demonstrate the defect in the diaphragm. It would be desirable to have a precise indication of the size and location of the defect, but this is not possible by present methods. The size of the hernia is an unreliable indication of the size of the defect in the diaphragm. This is more of theoretic than practical interest, however, as the indications for surgical intervention are clear. Even very

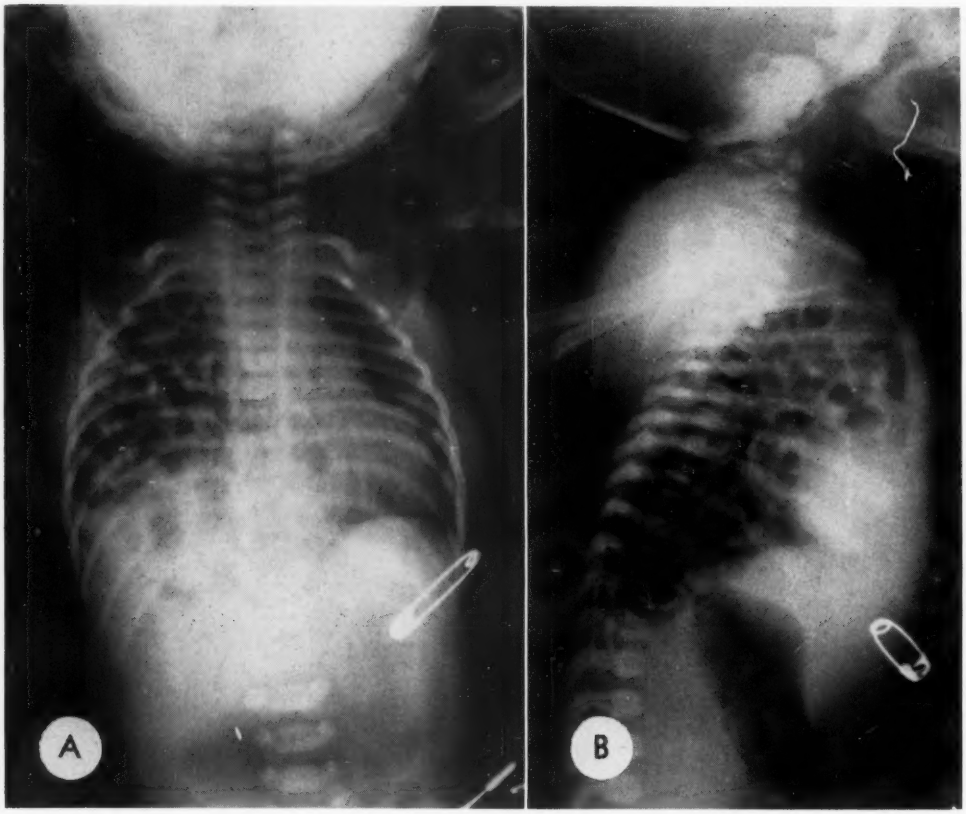


FIG. 4. (*A* and *B*) Right sided hernia in a thirty-six hour old infant with a 5 cm. defect involving the posterior aspect of the diaphragm. The patient recovered promptly after operative repair.

large defects in the diaphragm can be repaired in the newborn period by an experienced surgeon.

There is also uncertainty in the roentgenologic examination concerning the presence or absence of a hernial sac. That a sac is or is not present is determined by whether or not the herniation occurs before or after closure of the pleuroperitoneal membrane. If the abdominal contents extend freely into the thorax, the absence of a sac may be anticipated; whereas, if there is the appearance of a membrane confining the loops of intestine, the presence of a hernial sac may be expected.

The question is often raised as to whether a given situation represents a hernia with a hernial sac or an eventration involving the greater portion of the diaphragm or a small localized segment. While there is no doubt a marked difference in the opposite extremes of these conditions, it seems to us that there are instances where no real distinction can or should be made. A hernial sac is of diaphragmatic origin and is composed at least of its pleural and peritoneal surfaces. A localized eventration with absence of muscle in a portion of the diaphragm, on the other hand, tends to result in the formation of a similar sac. While there is often difficulty in making distinction by roentgenologic means, the problem is more academic than practical, as the indications for operation are largely clinical, and the experienced surgeon has techniques for dealing with either of these conditions.

The roentgen examination has an important role in the recognition and management of postoperative complications. The most obvious of these is the collection of air or exudate in the pleural space following either the thoracic approach or the abdominal approach when there is no hernial sac or when the sac has been opened.

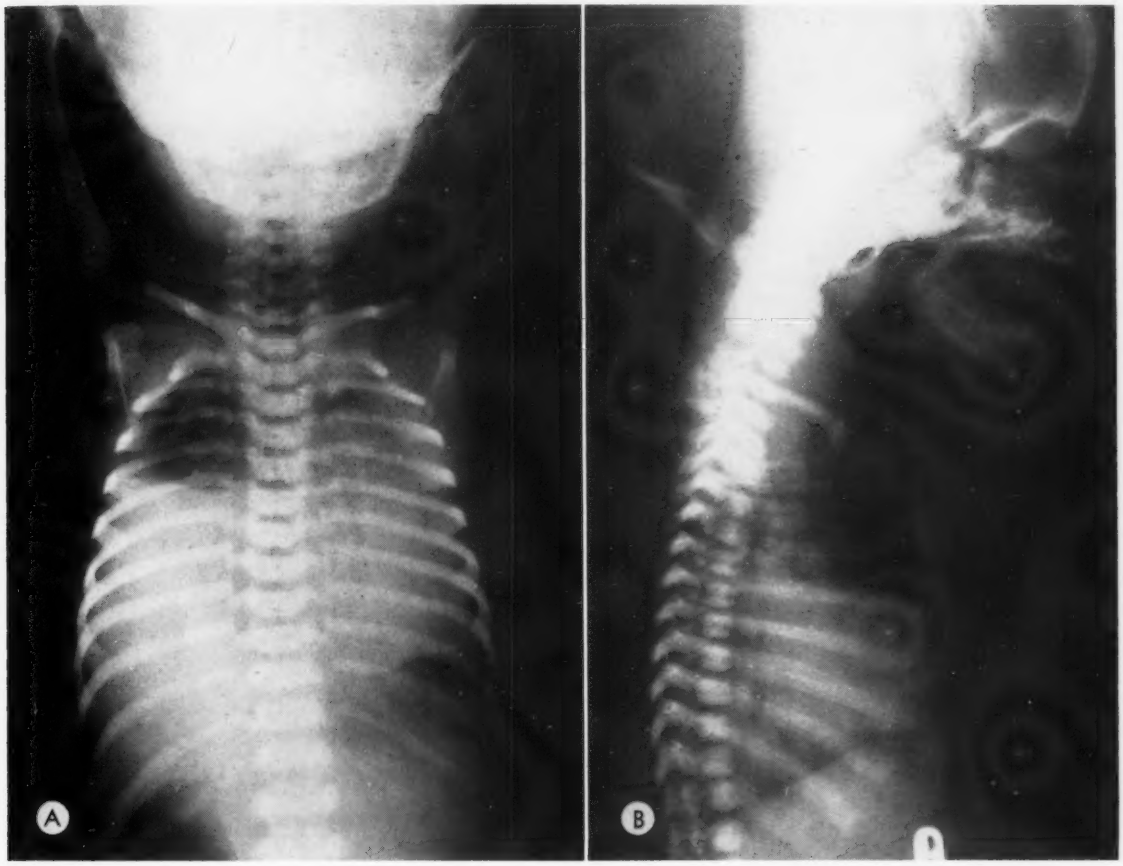


Fig. 5. (*A* and *B*) Massive density involving lower one-half of left hemithorax in a day old infant. Heart shifted markedly to the right. Death occurred at seven days of age. Autopsy revealed a 3 cm. defect in the posterior portion of the left diaphragm with herniation of the entire small bowel, cecum, ascending and transverse segments of colon, left kidney, adrenal gland and tail of pancreas.

Roentgenograms may be expected to give a reliable indication of the expansion of the lung. In some patients, when complete removal of the hernial sac was not feasible or when the sac was not recognized, it persisted as a cavity filled with fluid and gas in the thorax (Fig. 8, *A* and *B* and Fig. 9, *A*, *B*, *C* and *D*). These cavities slowly disappeared over a period of months and the patients were essentially asymptomatic. Tension pneumothorax was a common condition following abdominal approach (Fig. 10, *A* and *B*).

There still exists considerable controversy between advocates of the abdominal, the thoracic or the combined approach. Many patients in our series were operated from below. Whether or not the pneumothorax is associated with pulmonary hypoplasia, we are not prepared to state.

In a certain number of cases the problem of recurrence or incomplete reduction arises, and the roentgen study is very important in this situation. The radiologist must be aware of the possibility of a residual sac, as has been illustrated, and differentiation between this and a recurrence is sometimes quite difficult. Contrast study of the intestine should unhesitatingly be done in any doubtful situation (Fig. 11, *A*, *B*, *C* and *D*). There are 4 recurrences in this series and postoperative obstruction was encountered twice—both times due to incarceration in a residual or recurrent defect.

It is well recognized that the reduction of

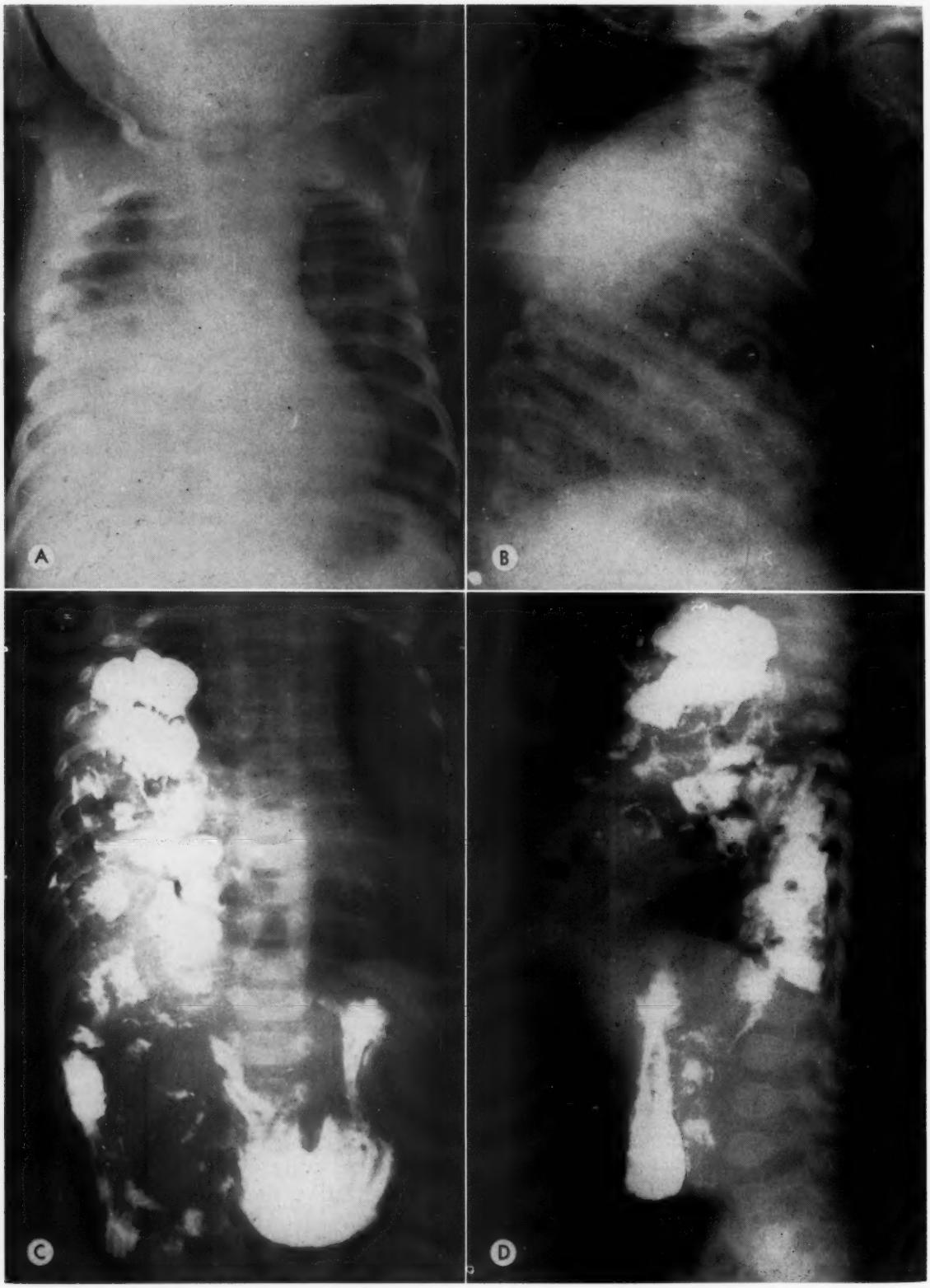


FIG. 6. (*A* and *B*) A right diaphragmatic hernia obscured by or misinterpreted as a pneumonic infiltration at the initial examination. (*C* and *D*) Diagnosis established three weeks later by following the course of barium administered by mouth. Reduction and repair was brought about through a thoracic incision at six months of age. Recovery was eventually complete although complicated by slow expansion of the right lung and manifestations of bowel obstruction.

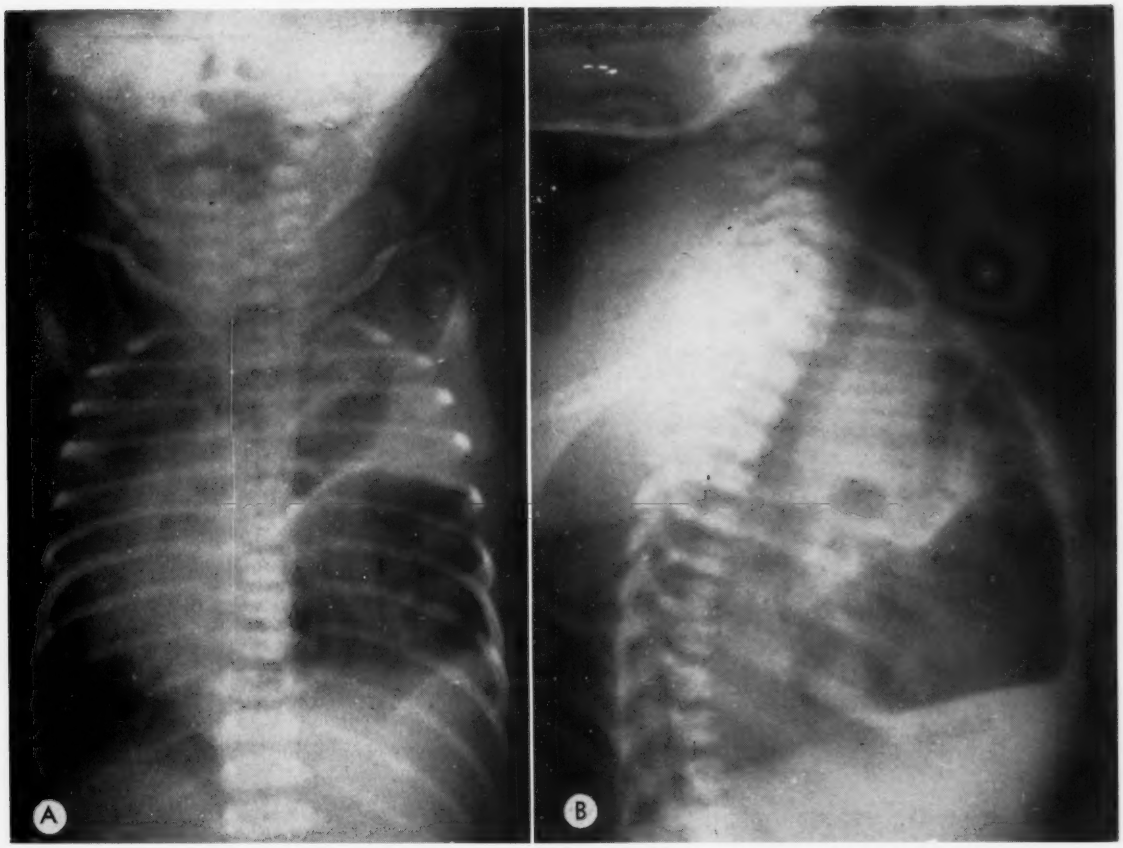


FIG. 7. (*A* and *B*) A large left sided hernia in a day old infant who expired within thirty minutes of admission Note the almost complete absence of aerated lung. Similar to Figure 4.

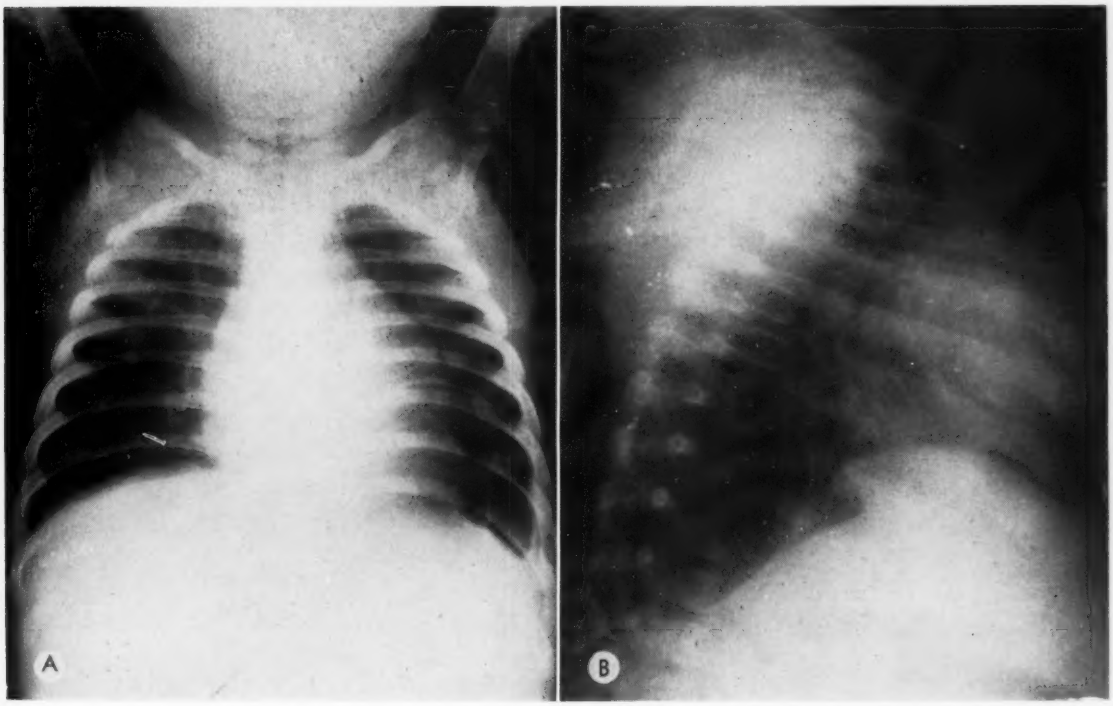


FIG. 8. (*A* and *B*) Cystic lesion observed in the lower lobe of the left lung from three to seven months following reduction of a hernia and repair of a left diaphragmatic defect in a two day old infant. The lesion was asymptomatic and was not demonstrated at an examination made at thirteen months of age.

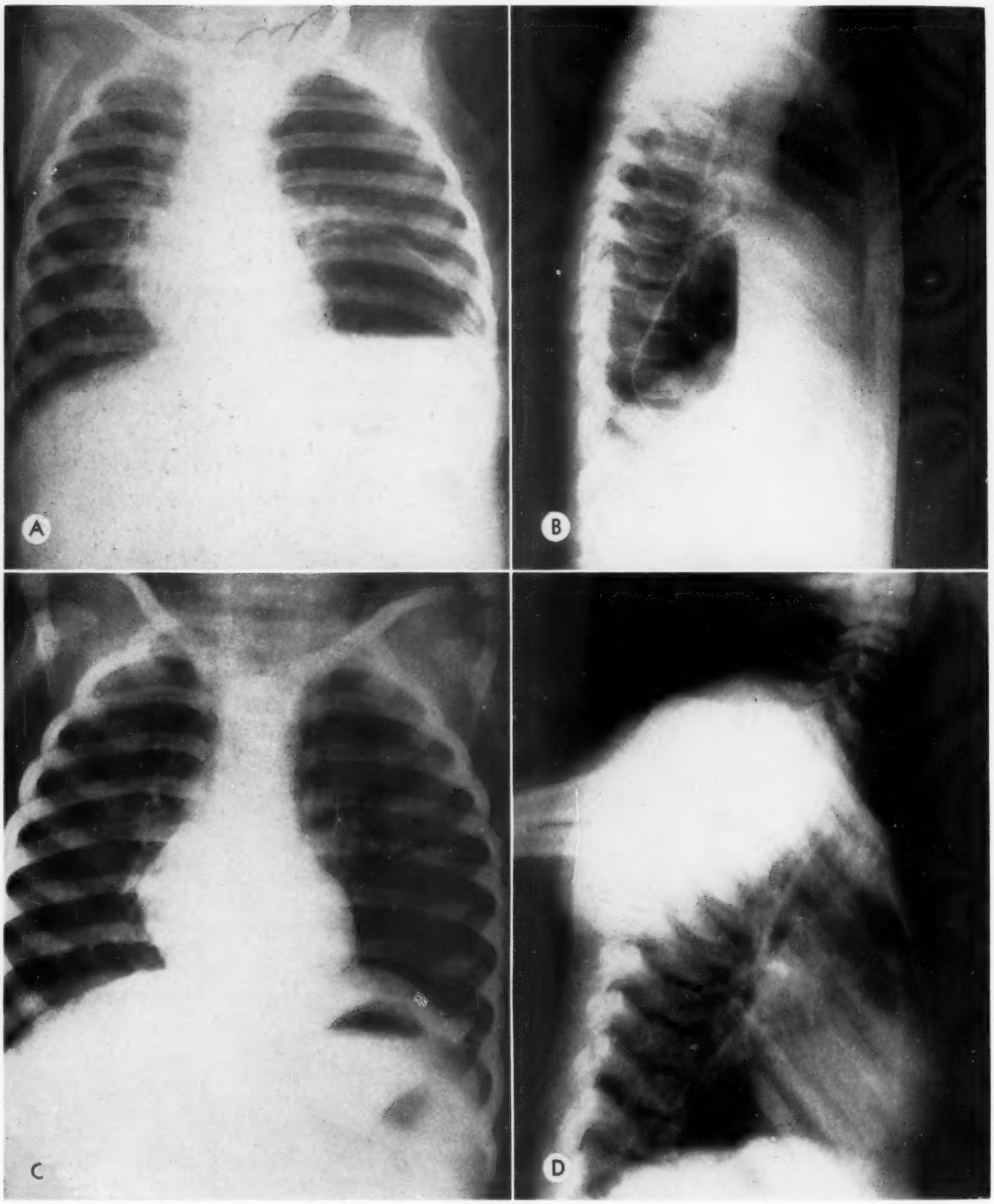


FIG. 9. (*A* and *B*) Postoperative chest roentgenograms demonstrating fluid-filled, cystic-appearing structure. (*C* and *D*) This structure disappeared after several weeks. It was believed to represent the hernial sac.

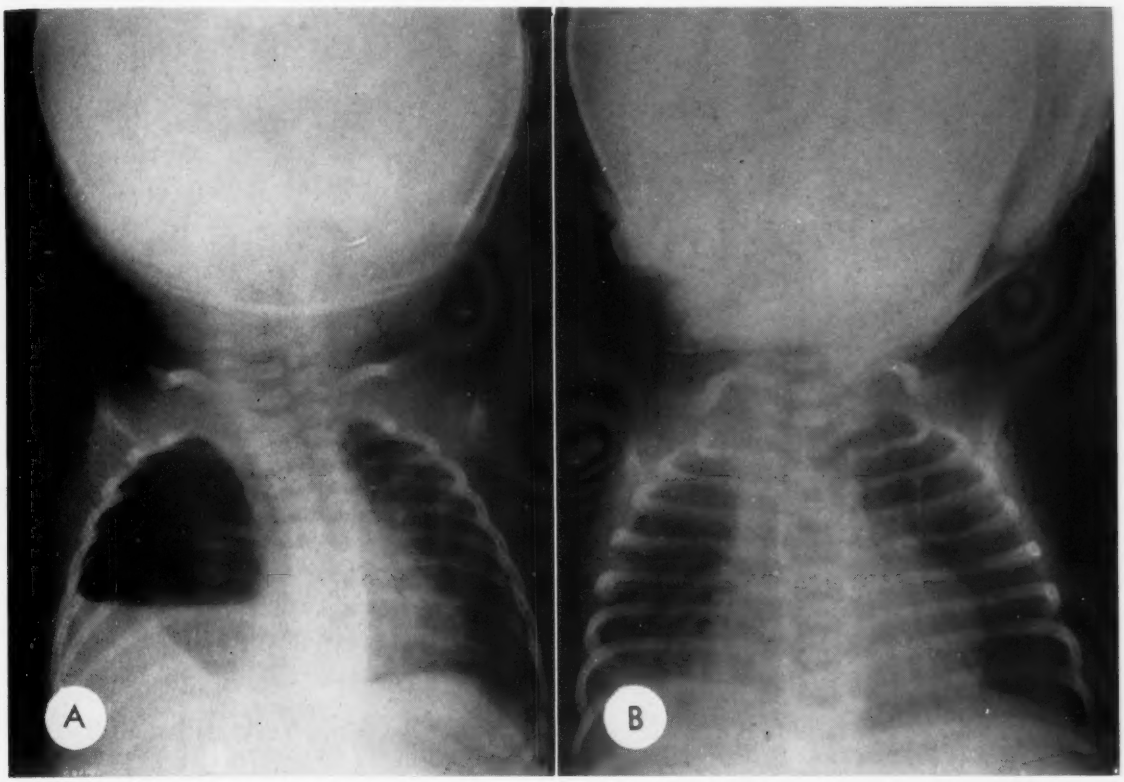


FIG. 10. (*A*) Postoperative roentgenogram of a repaired right-sided diaphragmatic hernia demonstrated a tension pneumothorax. (*B*) Complete re-expansion of lung followed prompt thoracentesis, and the subsequent course was uneventful.

a large hernia into a small abdominal cavity may give difficulty in the closure of the abdominal incision; or, if this is accomplished, there may be manifestations of intestinal obstruction from the tight abdominal wall. With the failure of proper mesenteric attachments that must be associated with a large congenital hernia, a situation is present following reduction which would seem to be favorable to the occurrence of volvulus. This has not been observed in our experience however, nor has it apparently been reported in the literature.

GROUP II. ESOPHAGEAL HIATUS HERNIAS

The frequent occurrence of these hernias in older people in contrast to their rare occurrence in infancy would indicate that these hernias are usually acquired. While some of these can be identified on plain roentgenograms of the chest, as in adults, all require contrast studies for satisfactory classification. They are usually found in the investigation of a feeding problem and recognized by the identification of a portion of the stomach lying above the diaphragm (Fig. 12, *A* and *B*). These are usually seen in the older infant; we have seen none large enough to interfere with life, nor severe enough to require immediate examination. Only one in this group of 12 was large enough to cause significant embarrassment of respiration (Fig. 13, *A* and *B*). This was of the para-esophageal type.

The roentgen diagnosis of this group presents the same difficulties as does diagnosis in adults. Variability in the size of the herniated mass, variation in the mucosal, muscular and serosal relationships, and difficulty in identification of the junction of the gastric and esophageal mucosa all

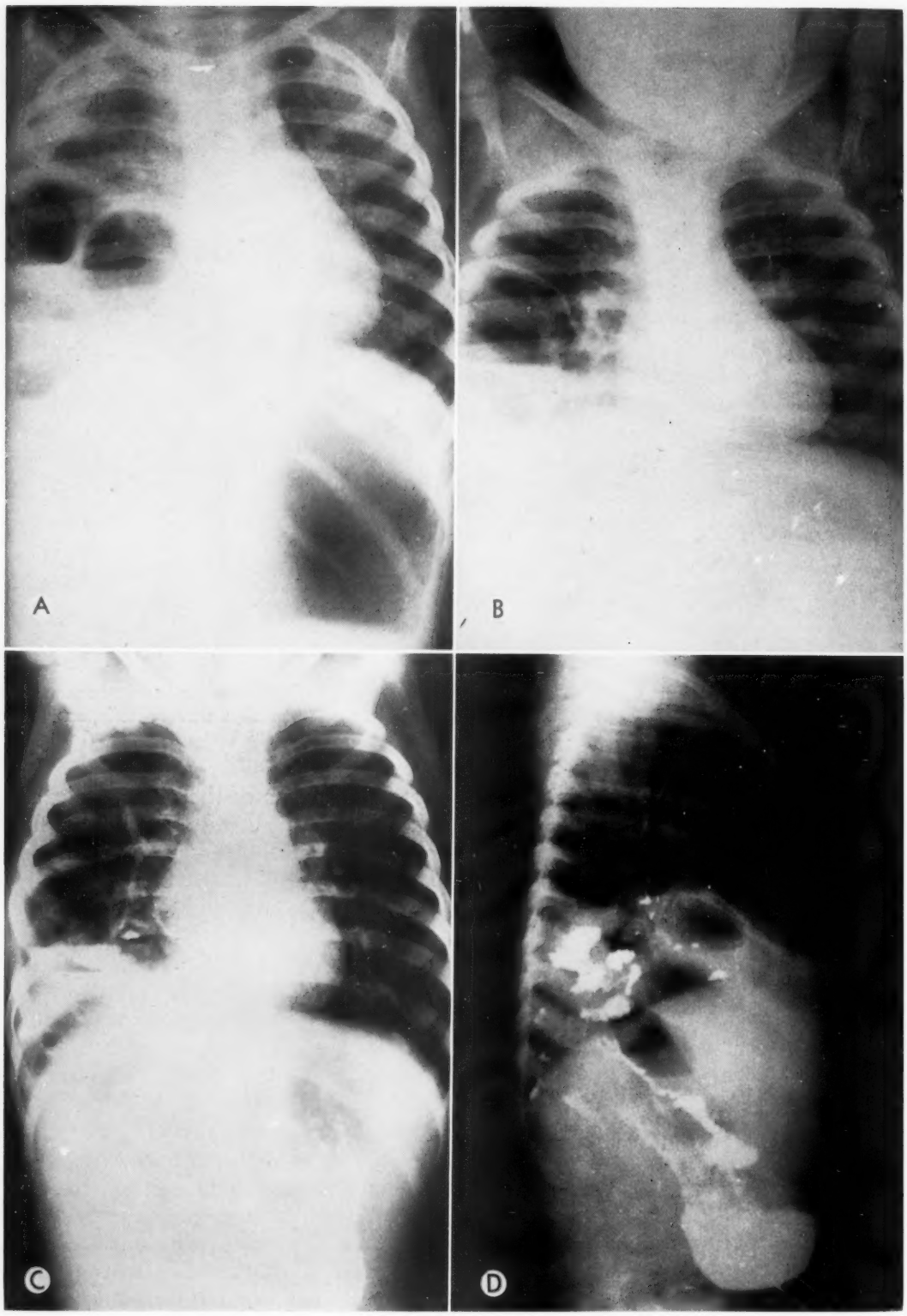


FIG. 11. (*A*) Roentgenogram made on the second postoperative day shows two large gas-filled structures with fluid levels. (*B*) Ten days later the configuration has changed somewhat, and subsequent gastrointestinal roentgenograms (*C* and *D*) showed the presence of incomplete reduction or recurrence of the diaphragmatic hernia.

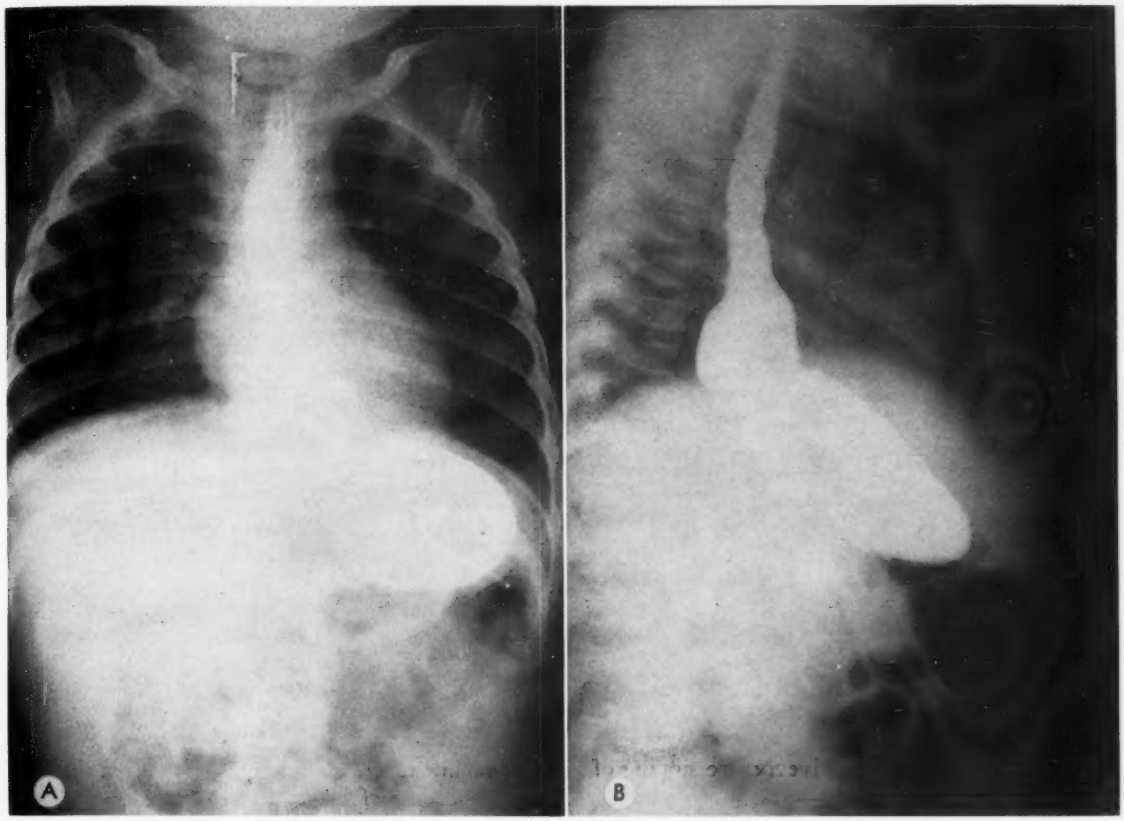


Fig. 12. (*A* and *B*) An esophageal hiatus hernia with shortened esophagus and probable ulcer on the posterior wall of the distal esophagus.

present problems. These problems are shared by the esophagoscopist and the anatomist.

### GROUP III. HERNIAS INVOLVING THE ANTEROMEDIAL ASPECT OF THE DIAPHRAGM

These hernias are sometimes referred to as occurring through the foramen of Morgagni which, like the foramen of Bochdalek, is a structure of doubtful reality. They may more properly be considered as hernias occurring in defects in the tendinous portion of the diaphragm, derived embryologically from the septum transversum. Often these defects assume the nature of eventrations. It has been our experience that mild degrees of eventration of the anteromedial segment of the right leaf of the diaphragm are so common as to be regarded as no more than a minor anatomic variation in development. No sharp distinction can be drawn between these insignificant eventrations and those larger ones that arouse speculation in diagnosis and treatment. From the reports in the literature of patients who have been operated upon, it would appear that the upper border of the liver conforms to the bulge in the diaphragm, and this nodule of the liver constitutes the content of eventration or hernial sac. No instances of this mild form of eventration are included in this report.

The situation in this group was much the same as in adults. The children were usually seen some time after birth, and the complaints arose because of the mechanical problems caused by herniation (Fig. 14, *A* and *B*).

### CONCLUSION

Emphasis is placed on the important factors in pleuroperitoneal hernia, which is the most common type of diaphragmatic hernia, and also the type most dangerous

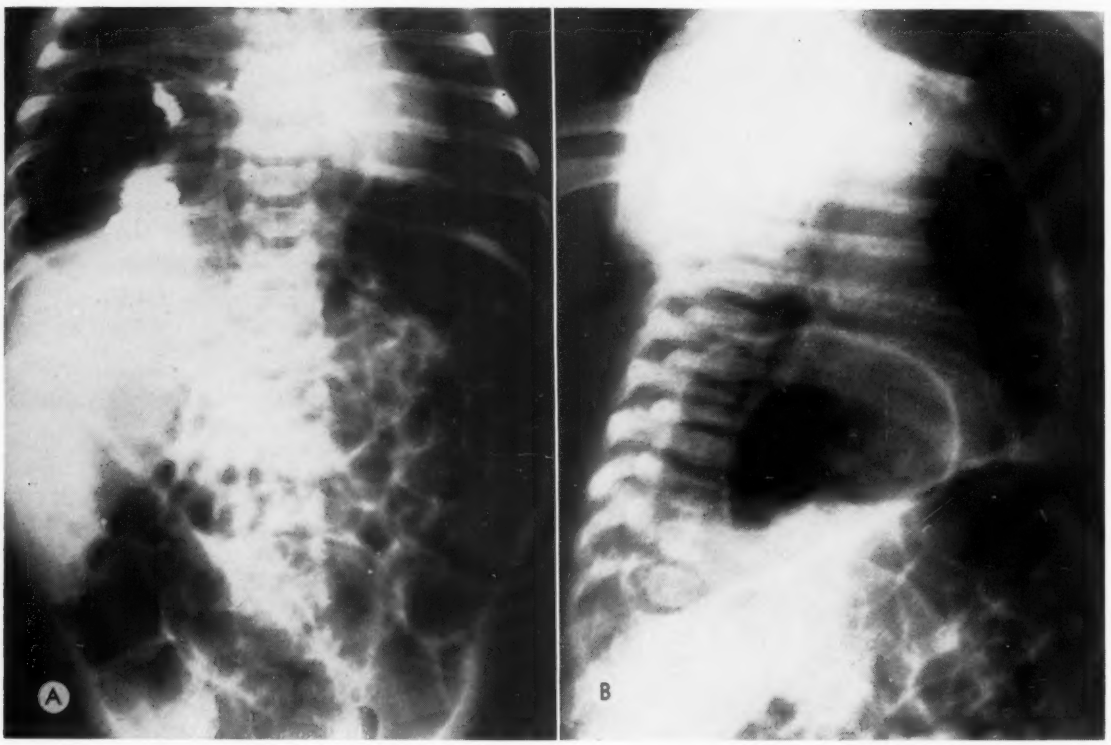


FIG. 13. (*A* and *B*) Preoperative roentgenograms of a month old infant who on two occasions became cyanotic after eating. Thoracotomy revealed a large para-esophageal type of hiatus hernia. The recovery was relatively uneventful with no history of recurrence.

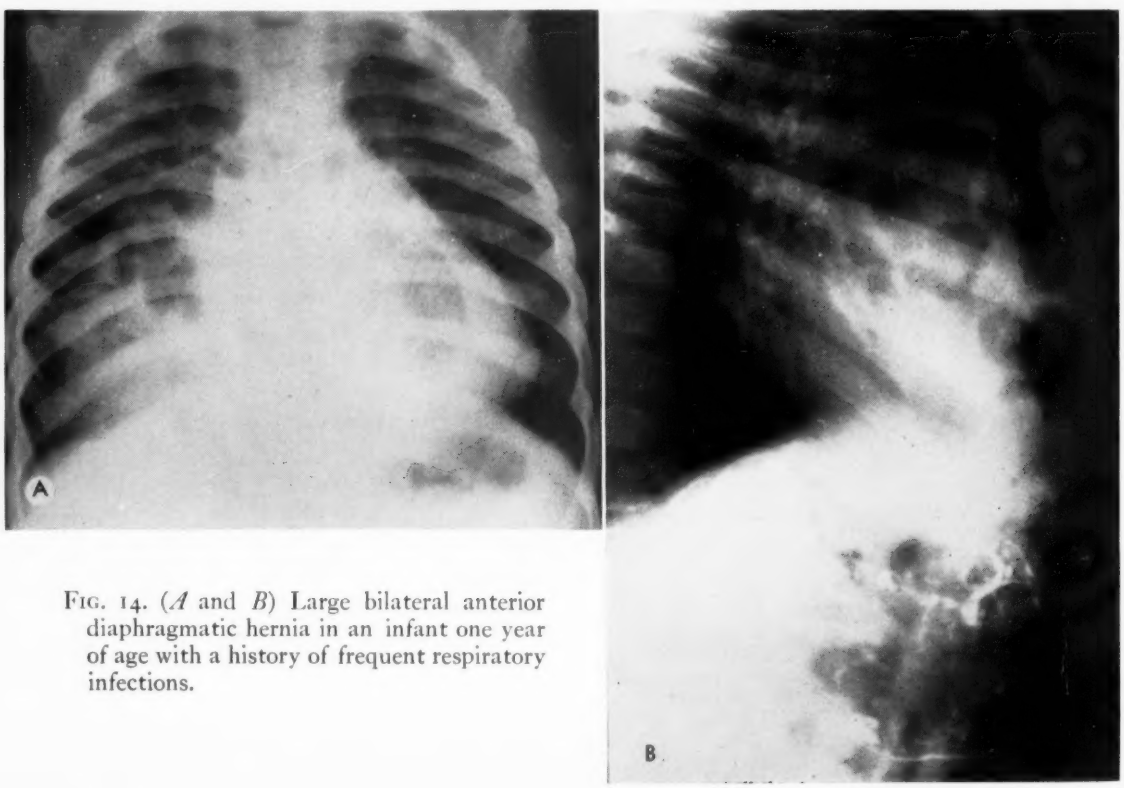


FIG. 14. (*A* and *B*) Large bilateral anterior diaphragmatic hernia in an infant one year of age with a history of frequent respiratory infections.

to the infant. The condition must be kept in mind as a possible basis for respiratory embarrassment. Plain roentgenograms are almost always diagnostic. A mass is demonstrated in the lower one-half to two-thirds of the hemithorax, usually showing fluid levels, displacement of the mediastinum to the opposite side, absence of a visible diaphragm on that side, absence of gas-filled intestine in the abdomen, and usually an esophagus of normal length. The possibility that the lung may be aplastic or hypoplastic rather than merely atelectatic should be considered.

J. O. Reed, M.D.
Children's Hospital of Michigan
Detroit 1, Michigan

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# DEFICIENCY OF SMALL INTESTINAL GAS IN INFANTS SIMULATING OBSTRUCTION

*By* HERBERT H. BENSON, M.D.,* *and* GEORGE JACOBSON, M.D.†
LOS ANGELES, CALIFORNIA

DEFICIENCY of gas in the small intestine of the newborn or infant is usually regarded as a cardinal sign of intestinal obstruction. Recently, however, Margulis and his co-workers[2,6] in several publications have called attention to the fact that this is not necessarily due to obstruction, but may be associated with adrenal insufficiency, dehydration, diarrhea, or may be seen where there is interference with normal transport mechanisms. The following cases are described to further emphasize these observations and to suggest the value of a radiopaque meal in differentiating true obstruction from simulated obstruction in early life.

## REPORT OF CASES

CASE I. A male infant, the first of premature twins born after an estimated gestation of seven months, was delivered in poor condition with cyanosis and marked respiratory difficulty. The birth weight was 2 pounds. The respiratory rate was 110 per minute and the cry was feeble. There was slight improvement in the cyanosis after treatment with oxygen.

A chest roentgenogram, taken approximately six hours after birth, showed pulmonary changes suggestive of hyaline membrane disease. There was also widening of the right upper mediastinum which was thought to represent atelectasis. A roentgenogram of the abdomen disclosed a small amount of gas in the stomach, but none in the small intestine or colon (Fig. 1).

The infant remained in critical condition and expired twelve hours after birth. The cause of death as determined at autopsy was massive intracerebral hemorrhage with dissection into the brain stem; and bilateral atelectasis. The gastrointestinal tract was normal with no evidence of malrotation, atresia, stenosis or obstruction.

CASE II. This female infant was the sibling of the infant described in Case I and was born shortly after its twin. The infant presented as a double footling breech and was delivered without difficulty in poor condition with a weak cry, shallow irregular respirations, and marked subcostal retractions. The birth weight was 1 pound, 14 ounces. A roentgenogram of the chest and abdomen was taken approximately six hours after birth. A moderate amount of gas was present in the stomach and proximal small

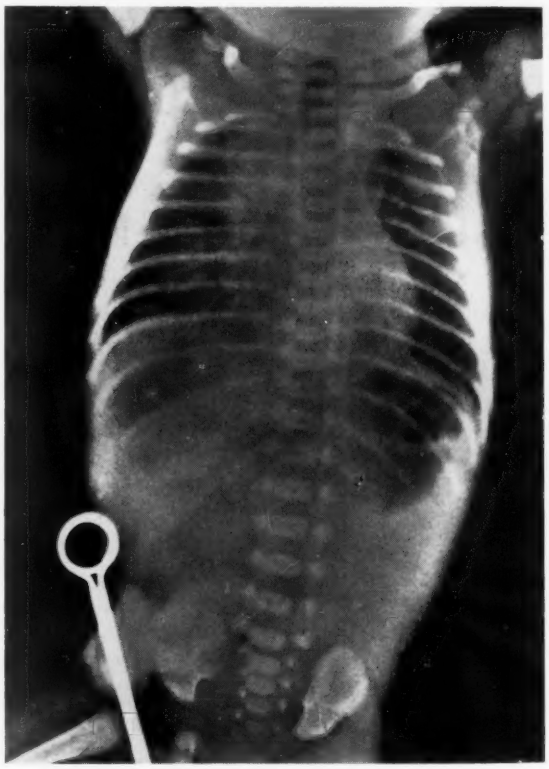

FIG. 1. Case 1. *Prematurity: Poor gas transport due to respiratory difficulty.* Roentgenogram taken six hours after birth shows a minimal amount of gas in the stomach. No gas is seen in the small intestine or colon.

* Resident in Radiology, Los Angeles County Hospital.
† Professor of Radiology (Head), Department of Radiology, School of Medicine, University of Southern California and Los Angeles County Hospital.

intestine, but none in the distal small intestine or colon (Fig. 2).

The infant remained in critical condition and expired ten hours after birth. The cause of death as determined at autopsy was prematurity and bilateral atelectasis. The gastrointestinal tract was found to be normal throughout with no evidence of obstruction.

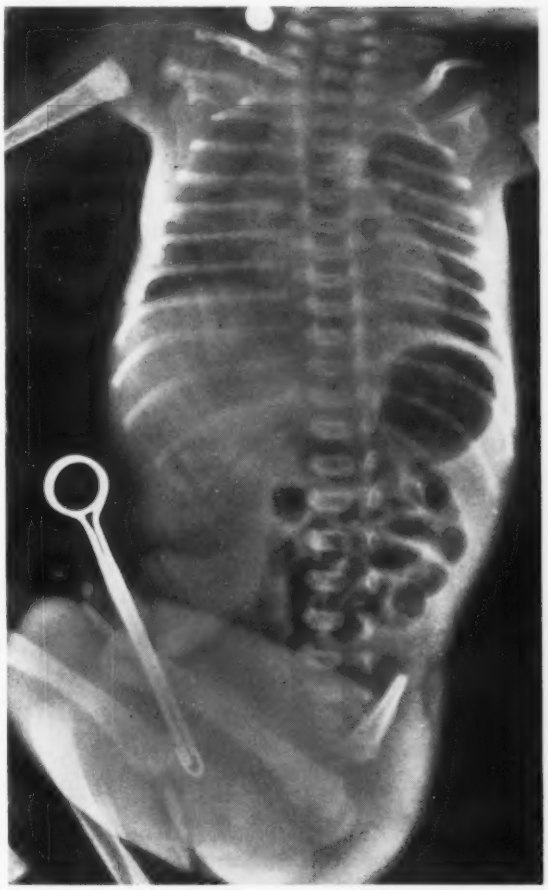

Fig. 2. Case II. *Prematurity: Poor gas transport due to respiratory difficulty.* Roentgenogram taken six hours after birth shows gas in the stomach and proximal small intestine but none in the distal small intestine or colon.

Case III. A ten day old female infant entered the Los Angeles County Hospital with a history of frequent emesis and black loose stools beginning seven days after birth. The infant was delivered normally at term and weighed 7 pounds, 9 ounces. For the first seven days of life regular feedings of Similac were given with breast feedings added on the third day of life. Stools were yellow and formed until the seventh day when they became black and loose, at which time the child began to vomit all food. The following day the stools became frankly bloody. On admission the stools were stated to be currant jelly in nature.

On physical examination the infant was found to be moderately dehydrated. The abdomen was soft, and peristalsis was hypoactive. The supine roentgenogram of the abdomen revealed a moderate amount of gas in the stomach with some additional gas bubbles which appeared to be in the small intestine. No other small intestinal gas or colonic gas was identified (Fig. 3*A*). The visible portions of the chest were not remarkable. A diagnosis of mid-gut volvulus with duodenal obstruction was entertained, with intussusception as a secondary possibility.

The infant was taken to surgery immediately and a laparotomy performed. There was no dilatation of the small or large intestine and all viscera were normal in position. The small and large intestine were examined from the ligament of Treitz to the rectum. No disease or abnormalities were identified, and the abdomen was closed. A roentgenogram taken the following day again disclosed only a small amount of gas in the stomach and in the proximal small intestine (Fig. 3*B*). The infant made an uneventful recovery and was discharged from the hospital ten days after admission.

The final diagnosis was nonspecific gastroenteritis.

Case IV. A five week old female infant was admitted to the Los Angeles County Hospital with a history of intermittent emesis, and diarrhea consisting of 5 or 6 loose green stools daily for the previous three days. The infant had a normal birth and weighed 7 pounds, 7 ounces at delivery. She had been on a formula of Carnation milk and water and had gained weight consistently until the onset of the present illness. On admission she weighed 8 pounds, 6 ounces and was moderately dehydrated. The abdomen was soft and the bowel sounds were hyperactive. Shortly after admission the infant passed several loose green stools, and approximately twelve hours after admission began vomiting "coffee ground" material.

A roentgenogram of the abdomen revealed an air-filled stomach of normal size and a large dilated loop of intestine in mid-abdomen (Fig. 4*A*). There were several small collections of air about this large loop which was thought to be

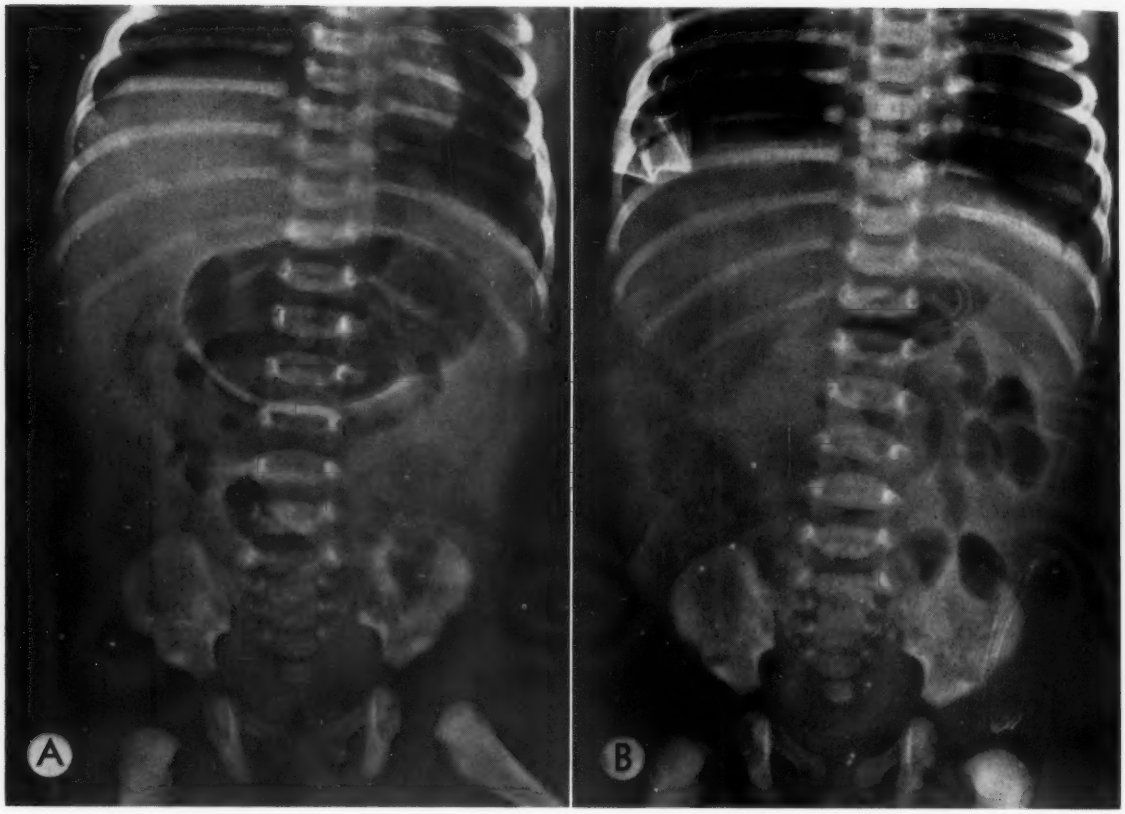


FIG. 3. Case III. *Nonspecific gastroenteritis.* (*A*) Roentgenogram taken on entry to hospital at age of ten days. The stomach contains a moderate amount of gas and there is only a small amount of gas in the proximal small intestine. (*B*) One day postoperative. There is still no visible gas in the distal small intestine or colon.

a dilated segment of the small intestine. Colonic gas could not be identified with certainty and there was no rectal gas present. An emergency barium enema examination was performed which disclosed a normal colon, and the previously dilated loop was confirmed as small intestine.

At this time an intestinal obstruction was considered and the infant was given a swallow of lipiodol. At the end of two hours the lipiodol was in the ascending colon and after four hours it was in the rectum (Fig. 4*B*). There was no evidence of obstruction. Subsequent treatment included hydration and antibiotics.

The infant made an uneventful recovery and was discharged seven days after admission. The final diagnosis was nonspecific gastroenteritis.

## DISCUSSION

The roentgenographic appearance of the normal infant abdomen differs primarily from that of the normal adult abdomen in that small intestinal gas is consistently present. Several investigators have shown that the normal infant swallows air with its first respiratory effort after birth. Fifteen minutes after birth, air has usually reached the jejunum and is distributed throughout the small and large intestine during the next two or three hours.[1,4,5,8] The transport time of intestinal gas is said to be fairly constant except in feeble or premature infants, in whom it may be lengthened.[4,9] Ordinarily, a considerable amount of small intestinal gas may be seen in children up to the age of four or five years. Absence or dearth of small intestinal gas in infants or young children, therefore, constitutes an abnormal situation which has almost always been interpreted as indicative of obstruction. Feinberg, Margulis and Nice,[2] however, have pointed out that the gasless abdomen in infancy may be due to a

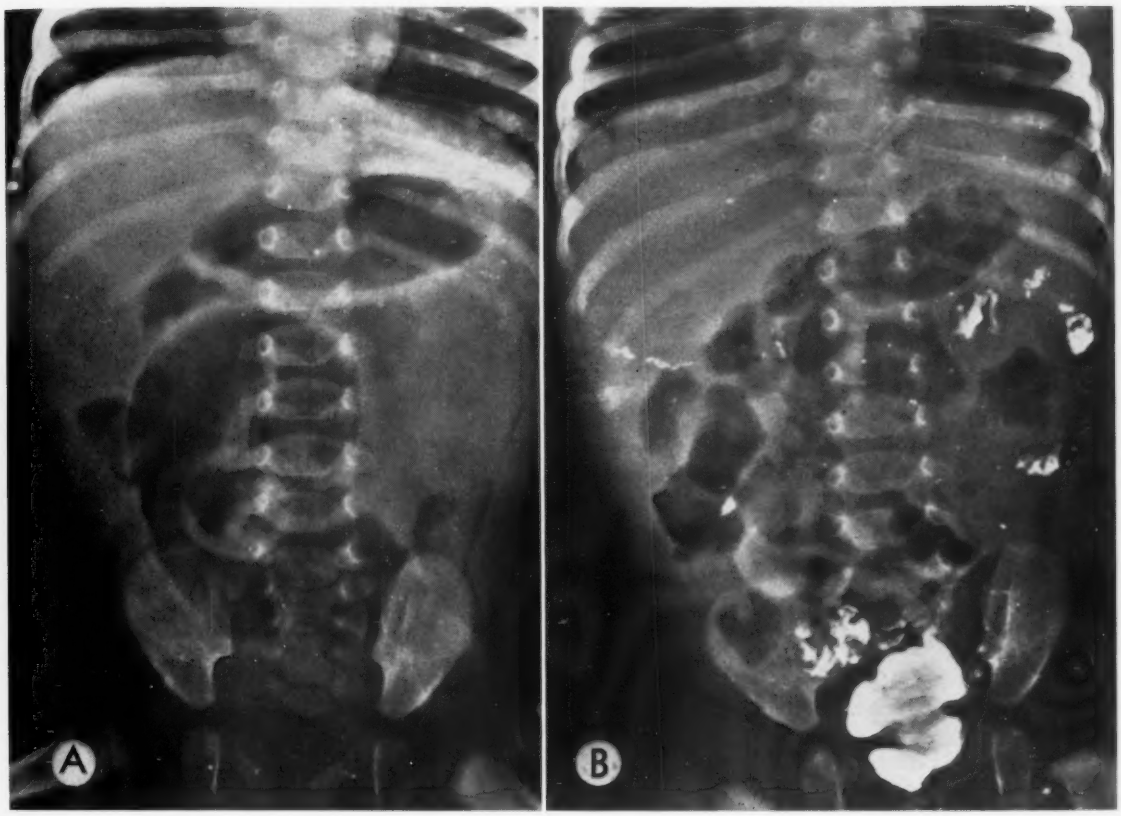


Fig. 4. Case IV. *Nonspecific gastroenteritis.* (*A*) Roentgenogram taken on entry to hospital at age of five weeks. The stomach contains a small amount of gas. A dilated loop of small intestine is seen in the mid-abdomen as well as some scattered small intestinal gas. No colonic gas is seen. (*B*) Four hours after oral administration of lipiodol. The contrast material is seen in the rectum excluding a mechanical obstruction. The small intestinal and colonic gas is still deficient in amount.

variety of causes, prominent among which are interference with normal transport mechanisms, dehydration, and diarrhea.

Respiratory effort was feeble in Cases I and II and consequently very little air was swallowed. Roentgenograms of the abdomen of both patients showed almost complete absence of small intestinal gas six hours after birth. Both infants were premature, both had atelectasis and, in addition, in Case I there was an extensive intracerebral hemorrhage.

Clinically, Case III was considered to be a small bowel obstruction. Roentgenograms could not exclude this possibility and surgery was performed which disclosed no evidence of obstruction or other abnormality. Stool cultures before and after surgery were negative for specific pathogenic organisms and it was concluded that the almost complete absence of small intestinal gas was due to a nonspecific enteritis. Case IV presented a similar clinical problem and the roentgenographic appearance of the abdomen was consistent with an obstruction. It was felt, however, that a contrast study might give further information and surgery was deferred. Lipiodol was given orally and rapid passage through small and large intestine demonstrated a patent gastrointestinal tract. Since the infant recovered rapidly on hydration and antibiotics, it was concluded that enteritis was responsible for the roentgenographic appearance of obstruction.

The use of contrast material given orally with suspected obstruction of the small intestine has been suggested by many in-

vestigators and appears to be a safe procedure. Barium, lipiodol and water soluble contrast media have been frequently used by us in adults with no unfavorable results. However, in infants we prefer not to use barium. Neither the oily nor water contrast media outline the mucosal structure as satisfactorily as barium, but patency of the intestinal lumen can be determined without difficulty. The water soluble media have an additional advantage; namely, hypertonicity which results in rapid passage through the small intestine facilitating early diagnosis. There is little or no absorption from the gastrointestinal tract and no toxic symptoms have been observed.

SUMMARY

Four cases are presented in which absent or diminished small intestinal gas in infants simulated obstruction but which proved to be due to respiratory difficulty in 2 cases, and dehydration and diarrhea in the other 2 cases. The value of oral contrast material in differentiating true from simulated obstruction is discussed.

George Jacobson, M.D.
1200 North State Street
Los Angeles 33, California

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# ROENTGENOLOGIC ASPECTS OF HYDROSTATIC REDUCTION OF ILEOCOLIC INTUSSUSCEPTION*

*By* BERTRAM R. GIRDANY, M.D., LEE W. BASS, M.D., *and* WILLIAM K. SIEBER, M.D.
PITTSBURGH, PENNSYLVANIA

HYDROSTATIC reduction by barium enema is now an accepted method of treatment of ileocolic intussusception.[2] It diminishes the morbidity associated with uncomplicated ileocolic intussusception.[1] No significant differences in mortality exist between hydrostatic and primarily surgical methods of reduction. Wider use of the roentgen method allows earlier identification of intussusception in suspected cases and obviates unnecessary surgery associated with errors in diagnosis.

Hydrostatic reduction of ileocolic intussusception has been the initial treatment of choice at the Children's Hospital of Pittsburgh for seven years, and has been used in 106 cases. This paper considers the technical problems associated with the hydrostatic procedure. It presents criteria for completeness of reduction of ileocolic intussusception, gives roentgenographic examples of lead points and ileo-ileal intussusception, and discusses causes of failure of hydrostatic treatment.

## LITERATURE AND DISCUSSION

This paper does not argue the merits of hydrostatic pressure as opposed to laparotomy in the treatment of uncomplicated ileocolic intussusception. Ravitch and McCune,[7,8] Bass, Sieber and Girdany,[1] Nordentoft,[4,5] and Nordentoft and Hansen,[6] have discussed this problem. The hydrostatic method lowers patient morbidity, does not increase mortality rates, is not associated with increased rates of recurrence, and does not dispose to perforation of bowel. Bass *et al.*[1] emphasize clinical factors that contribute to poor prognosis. They indicate that duration of disease is not a necessary contraindication to hydrostatic pressure, and that any method of reduction of intussusception is contraindicated until dehydration and shock have first been corrected.

The radiologist applying hydrostatic treatment to a child with ileocolic intussusception must be familiar with the clinical condition. Intussusception reflects invagination of bowel into its own lumen, leading into the lumen of distal bowel. The term "ileocolic" refers to intussusception beginning in the distal ileum and leading into the lumen of proximal colon. "Ileo-ileal" indicates origination of the intussusception at any site in the ileum proximal to its terminal portion. "Ileo-ileocolic" refers to ileo-ileal intussusception that has progressed to involve the colon. Because the clinical behavior of the two latter types of intussusception is similar, the conditions are usually considered together. The term "uncomplicated" refers to ileocolic intussusception not caused by a basic anatomic abnormality or "lead point," and not associated with gangrene.

Intussusception is a dynamic condition; the head of the intussusception is not in fixed position unless or until gangrene and adhesions occur. Stretch on the mesentery produces periodic shock-like episodes and circulatory embarrassment, the latter resulting in passage of blood in the stool. Gangrene may be an early or late occurrence, depending on the extent of vascular embarrassment.

Ileocolic intussusception is an idiopathic condition usually occurring in previously healthy children between the ages of three months and three years. There is no regular antecedent history; it is heralded by attacks of severe paroxysmal abdominal pain

* From the Departments of Radiology and Surgery, Children's Hospital of Pittsburgh and the University of Pittsburgh, School of Medicine.

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FIG. 1. Roentgenogram shows the characteristic filling defect in the barium column at the site of the intussusception in the proximal portion of the transverse colon.

associated with pallor. Vomiting, palpable abdominal mass and "currant-jelly" stools may be present. Any or all of these clinical findings may be absent in the individual case. Roentgenologic examination of the colon with opaque medium affords certain identification of ileocolic intussusception (Fig. 1).

Plain roentgenograms of the abdomen may be useful in identification of intussusception, but are unreliable. The gas and soft tissue pattern in the abdomen is often normal. Absence of air in the region of the cecum and ascending colon is an inconstant finding. Signs of intestinal obstruction may contraindicate hydrostatic treatment and indicate the need for surgical exploration. They suggest the presence of gangrene and/or ileo-ileal intussusception (Fig. 2). Eight of the 11 children with ileo-ileal intussusceptions in this series showed roentgenographic signs of intestinal obstruction. In addition, 7 children with roentgen changes of intestinal obstruction had ileo-ileal intussusceptions and were not offered hydrostatic treatment.

The child's clinical condition is evaluated; shock and dehydration are corrected before any definitive treatment of intussusception is undertaken. After the surgeon has been consulted and is in attendance, the radiologist applies hydrostatic pressure with barium enema. Normal saline solution, made by the addition of a level teaspoonful of table salt to a pint of lukewarm tap water, is used as the vehicle for the opaque material to eliminate the possibility of water intoxication. A thin mixture of micropaque or other colloidal suspensions of barium allows adequate contrast visualization, prevents settling of the barium and eliminates the need for constant agitation of the solution in the enema reservoir. The examiner tests the patency of the fluid system by observing the free flow of the barium mixture before he inserts the enema tube.

The radiologist protects himself and his assistants from exposure to radiation, and limits radiation exposure to the patient. Immobilization of the patient is as important to the success of hydrostatic treat-

FIG. 2. Plain roentgenogram of the abdomen in a three year old child shows signs of small bowel obstruction. There was a palpable abdominal mass, and a history of paroxysmal abdominal pain. Hydrostatic treatment was not attempted. At surgery, ileo-ileal intussusception was identified and resection of gangrenous bowel was necessary.

ment as control of the flow of opaque material. The inadequately immobilized infant is for the radiologist the equivalent of the poorly anesthetized patient for the surgeon. The technique of immobilization has been described elsewhere.[3] It utilizes heavy sand bags, nonelastic Ace bandages, and compression bands (Fig. 3).

The radiologist introduces a nonlubricated Bardex type, No. 24 catheter into the infant's rectum and distends the bag with air. His choice of catheter depends on the size of the patient. A large lumen insures transmission of hydrostatic pressure. The distended balloon fits snugly against the perineum at the internal anal orifice. Further insertion of the tube results in kinking of its tip and interferes with free flow of the enema solution. Overdistention of the balloon occludes the lumen of the tube. Large strips of adhesive tape placed across the patient's buttocks binding them together help prevent expulsion of the tube from the rectum.

The enema reservoir is raised to a height of 3 feet. This is an arbitrary, but proved safe, height. Neither perforation nor reduction of gangrenous bowel occurs with 3 feet of hydrostatic pressure.[7,8] Using a small fluoroscopic field, the radiologist observes the retrograde passage of the opaque material through the colon. The enema tube must not be "milked" or compressed to promote the flow of solution. Complete intestinal obstruction produces a closed hydrodynamic system. "Milking" of the tube may produce a major increase in intraluminal pressure similar to that effected by the pressure of a foot on the brake pedal in an hydraulic brake system. The single perforation that occurred in this experience of 106 cases followed prolonged "milking" of the enema tubing after the flow of opaque material into the ascending portion of the colon had ceased. Perforation took place in normal colon and not in gangrenous bowel.

The reduction of intussusception may proceed slowly, particularly if the intussuscepted bowel has progressed distally

Fig. 3. Sand bags, stockinette pulled over the arms, and Ace bandage wrapped around the lower extremities immobilize the infant. A compression band may be drawn across the lower extremities. (Reproduced by permission from *Radiology*, 1953, *60*, 518–526.)

into the descending and sigmoid portions of the colon. Flow of the barium mixture is often slow at the hepatic flexure and through the ileocecal valve. It is not necessary for the radiologist to keep the head of the barium column in these areas under constant fluoroscopic observation. Prolonged radiation exposure to the patient must be avoided. Manual palpation of the patient's abdomen is not employed. Hydrostatic pressure alone provides the force pushing the displaced bowel back to its normal site.

Retrograde filling of the ileum begins at the ileocecal valve and proceeds proximally with the loops of barium-containing small bowel lying along the medial and proximal aspect of the ascending portion of the colon. The barium mixture then fills the loops of small bowel that lie along the superior and medial margins of the ascending colon and along the inferior and proximal margins of the transverse colon (Fig. 4). The radiologist recognizes completeness of reduction of ileocolic intussusception by observing the normal pattern of flow of opaque solution in the ileum.

In the absence of immediate reflux of opaque material through the ileocecal valve into the ileum, the radiologist waits for five minutes, during which period he makes occasional short fluoroscopic observations. He again checks the patency of the fluid

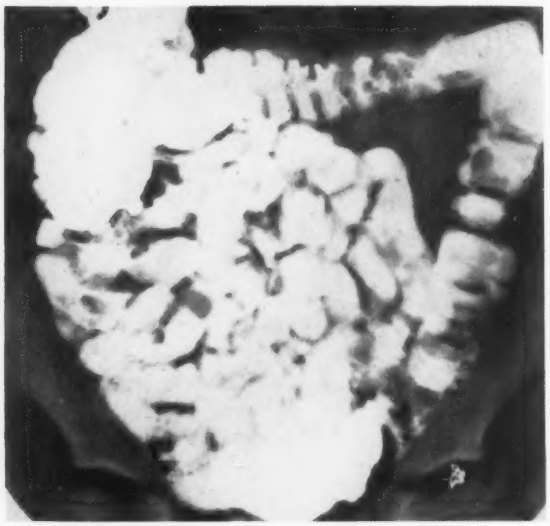

FIG. 4. The distribution of barium mixture in the small bowel after successful hydrostatic reduction of intussusception is seen.

system. Cessation of flow may follow depletion of solution in the reservoir, kinking of the tubing, or extravasation of the barium mixture on the table. If the ileum does not fill after the five minute period of waiting, the radiologist terminates the hydrostatic procedure. The infant is allowed to empty his colon, and a postevacuation roentgenogram of his abdomen is made. Defecation may produce reflux of barium mixture into the ileum.

Absence of reflux of opaque material into the ileum may occur despite complete reduction of intussusception. Nonetheless, ileal filling is the most important single roentgen criterion of completeness of reduction. Without this sign the radiologist cannot declare reduction complete; the attending surgeon then must decide the course he is to follow after observing the clinical progress of the patient. Edema of the ileocecal valve may be present after successful reduction, and may simulate the lead point of an incomplete reduction (Fig. 5). Localized filling defects in the ileal lumen indicate local abnormalities or lead points. Figure 6 shows the roentgenographic findings in a child with duplication of the ileum in whom reduction of ileocolic intussusception was achieved but a filling defect in the distal ileum persisted. Figure 7 shows the characteristic findings in ileo-ileal intussusception. Ileo-ileal intussusception is not treated hydrostatically; the probability of lead point is high, and hydrostatic pressure may not be well transmitted past the ileocecal valve. Seven of the 11 children with ileo-ileal intussusceptions had lead points demonstrated at surgery. Three other patients in this group had gangrenous segments of ileum.

Recurrent intussusception most commonly occurs within the twenty-four hour period after reduction by either hydrostatic or surgical means. A plain roentgenogram of the abdomen made within twenty-four hours after the application of the hydrostatic method is useful in the identification of recurrence of intussusception. Recurrence of pain is an indication for roentgen examination of the abdomen. The retained barium mixture may allow accurate identification of the recurrence (Fig. 8).

The treatment of recurrent intussusception is not within the scope of this paper.[9] Intussusceptions recurred in 8 patients. Five of these patients were operated upon. In 4, no obvious lead point was present and in the fifth there was hypertrophy of the lymphoid tissue in the wall of the ileum. Three infants had no further recurrences after a second reduction by hydrostatic pressure.

## RESULTS

Hydrostatic pressure with barium enema was the initial treatment of intussusception in 106 children. In 87 instances of uncomplicated ileocolic intussusception, this treatment was definitive. There were 5 failures of the hydrostatic method in the treatment of uncomplicated ileocolic intussusception. In these 5 instances reduction was completed by surgery at laparotomy. Two of these failures occurred in the first year that the hydrostatic method was used and may have been due to inexperience with the technique. One failure oc-

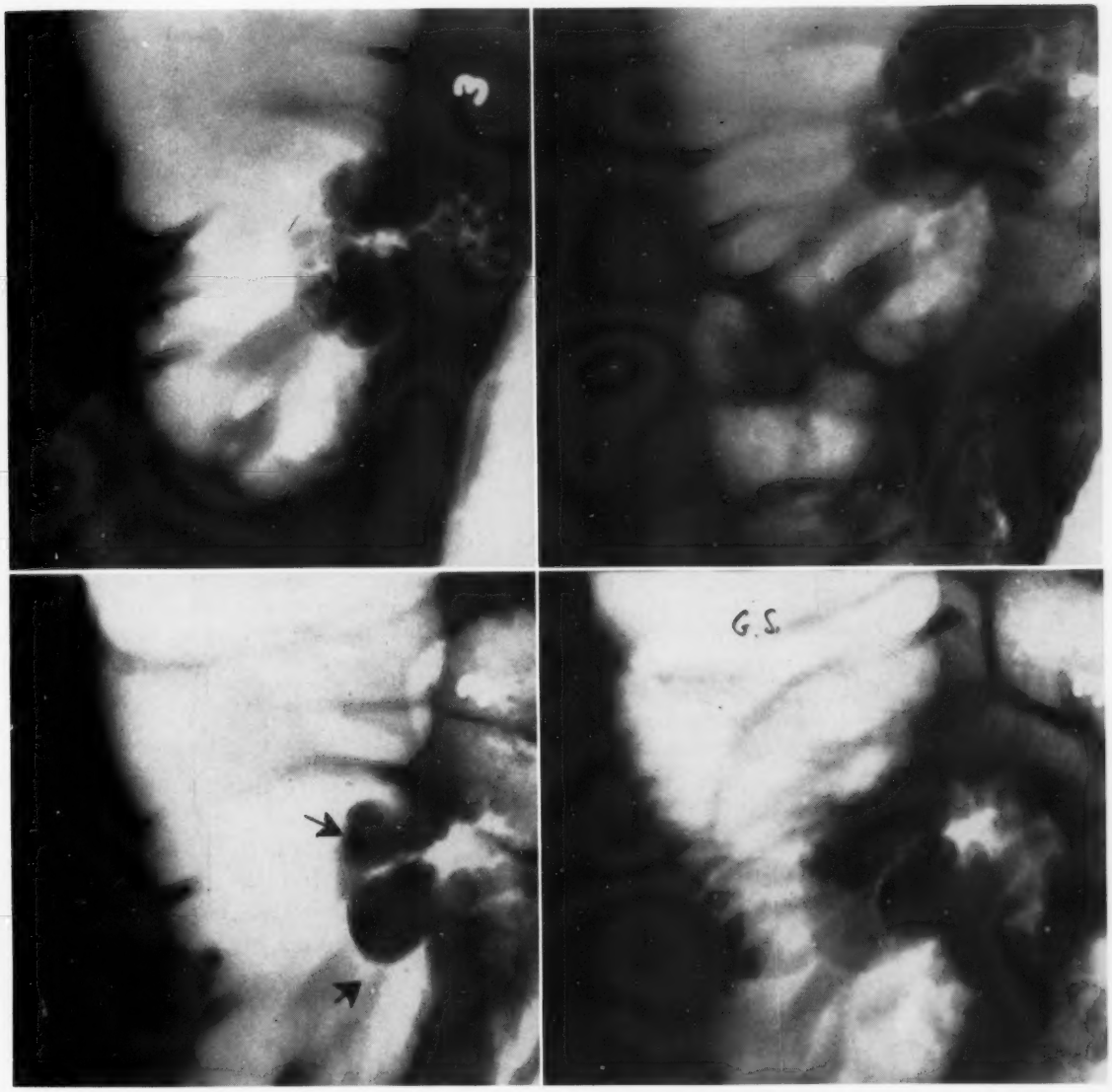


Fig. 5. Spot roentgenograms of the ileocecal area made during hydrostatic reduction of intussusception show persistence of filling defect at the ileocecal valve. In this instance, recurring intussusception led to surgical intervention. At surgery edema of the mucosa of the ileocecal valve was proved and was the probable cause of recurrence of intussusception.

curred in an intussusception that presented at the anus. Hydrostatic pressure slowly returned the intussuscepted bowel to the proximal portion of the ascending colon. At surgery reduction of the remaining portion of the intussusception was difficult. There were many adhesive bands, and the bowel was almost gangrenous. Hydrostatic pressure failed to reduce ileocolic intussusception in 1 infant with signs of intestinal obstruction. This infant, at surgery, had associated malrotation of the bowel. Ileoileal intussusception recurred following surgical reduction and re-operation was necessary. The fifth failure was associated with signs of intestinal obstruction. The postevacuation roentgenogram of the abdomen showed persistence of a large filling defect in the proximal colon, but reduction occurred spontaneously during administra-

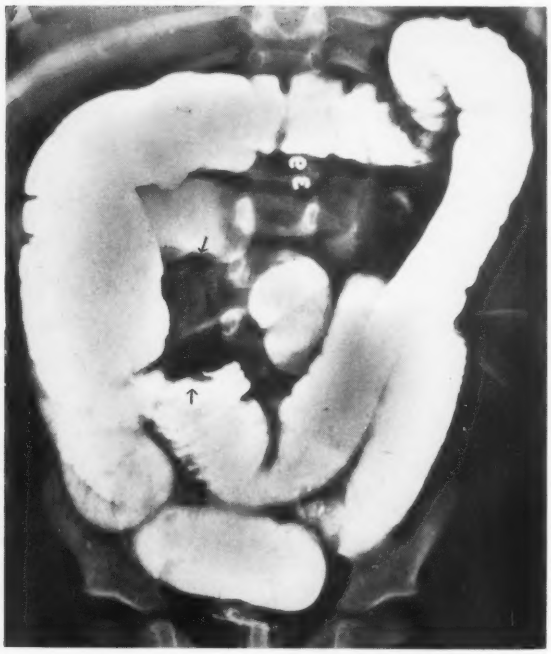

FIG. 6. The arrows are directed at a filling defect in the ileal lumen after hydrostatic reduction of intussusception. The filling defect was caused by duplication of the ileum which was resected surgically.

FIG. 7. The findings of ileo-ileal intussusception are seen.

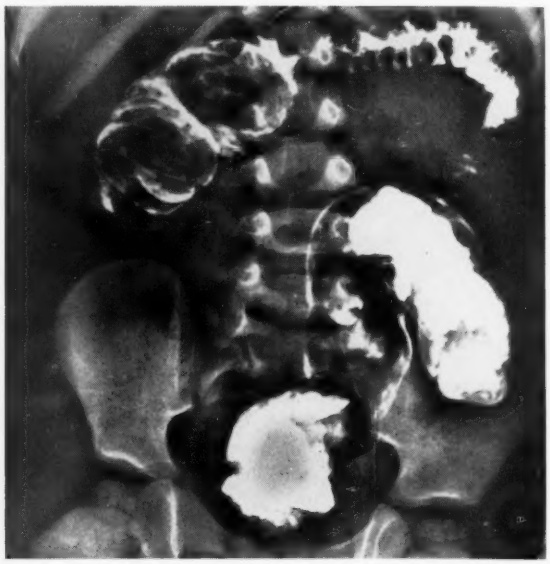

FIG. 8. Plain roentgenogram of the abdomen made twelve hours after successful hydrostatic reduction of ileocolic intussusception clearly shows signs of recurrent intussusception.

tion of anesthesia before laparotomy. At surgery the terminal ileum was "dusky."

Two children with ileocolic intussusception had gangrenous bowel at surgery which required resection. The single death in this series occurred two days after hydrostatic reduction of ileocolic intussusception. This infant was moribund on admission and the hydrostatic pressure with barium enema was used at the request of the attending surgeon because it seemed improbable that the infant would tolerate laparotomy. Necropsy showed complete reduction of nongangrenous bowel.

In 11 children with ileo-ileal intussusceptions hydrostatic treatment was undertaken. Eight of these patients had plain roentgenographic findings of intestinal obstruction. In 4 of the 11 children hydrostatic pressure failed to provide reflux of opaque material into the ileum. Of the other 7 patients with ileo-ileal intussusceptions, 3 had lead points roentgenographically identified as filling defects in the barium column in the ileum, and 4 showed characteristic signs of ileo-ileal intussusception. One infant in this group had complete reduction of the ileo-ileal intussuscep-

tion by hydrostatic means. The surgeon declined to operate because of the patient's subsequent clinical recovery.

## CONCLUSIONS AND SUMMARY

1. Roentgenographic aspects of hydrostatic reduction of ileocolic intussusception have been discussed.

2. Hydrostatic pressure with barium enema is an effective method of reduction of uncomplicated ileocolic intussusception.

3. Failure of successful reduction by hydrostatic pressure suggests the presence of ileo-ileal intussusception or gangrene.

4. Plain roentgenographic findings of intestinal obstruction are contraindications to the use of hydrostatic pressure and usually indicate ileo-ileal intussusception and gangrene.

Bertram R. Girdany, M.D.
Children's Hospital of Pittsburgh
Pittsburgh 13, Pennsylvania

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# ROENTGENOLOGIC MANIFESTATIONS OF GLIOMA OF THE OPTIC NERVE AND CHIASM*

*By* COLIN B. HOLMAN, M.D.
*Section of Roentgenology, Mayo Clinic and Mayo Foundation*†
ROCHESTER, MINNESOTA

THE diagnosis of glioma of the optic nerve or chiasm or both is of considerable practical importance. If discovered relatively early, this is one form of glioma in which the prognosis may be favorable.[5] Roentgenologic diagnostic methods are appealing because a large number of these patients are small children and, therefore, it may be difficult to obtain a reliable history of changes relative to vision and the ophthalmologic examination may be exceedingly difficult.

Since the paper by Martin and Cushing,[19] in 1923, several reports[1,7,10–12,17,23,24] have dealt to some extent with the roentgenologic features of these tumors. However, considerable variation exists in the manifestations noted in these studies. Many reports included different although somewhat similar lesions that occur in or near the same anatomic region. Others included cases in which the diagnosis was not proved pathologically. Furthermore, although intracranial tumors of children have been investigated, little emphasis has been placed on the relationship of a growing tumor to a growing sphenoid bone as a clue to the variability in the reported features.

Therefore, a study was made of all available roentgenograms in 48 patients seen at the Mayo Clinic who were proved to have these tumors in order to determine what features might be of diagnostic value. Only the roentgenologic manifestations will be considered, since the clinical, ophthalmologic, pathologic and surgical aspects in most of these cases have been reported previously.[5]

Gliomas of the optic nerves and chiasm usually are found in children or young adults. They grow slowly and sometimes are present at birth.[5] Thus, their roentgenologic manifestations depend to a considerable degree on the effect of the growing mass on the adjacent portions of a growing cranium. Therefore, it is useful to describe these changes according to the age of the patients affected, the site of origin of the tumor and its direction of expansion. The material has been analyzed with respect to younger children, older children and young adults, as well as with regard to tumors occurring only within the optic nerve and those involving the optic chiasm or both the nerve and chiasm.

## TUMORS OF THE OPTIC NERVE ALONE

These neoplasms may involve only the intra-orbital portion of the nerve. In such cases, no changes in the bony orbit or calvarium either in newborns or older patients were noted in this series. Proptosis sometimes may be indicated roentgenologically by bulging of the anterior soft tissues in the orbital region.

Most tumors involving the optic nerve lie, at least in part, within the optic canal of the sphenoid bone, and the roentgenologic changes concern this region, which includes the lesser wing of the sphenoid and the anterior clinoid process. These changes imply some intracranial extension of the tumor.

The optic nerve only was involved in 13 of this series of patients; 11 were female and 2 were male. In 9 of these patients, roentgenograms of the optic foramina employing the Rhese position were made; 7 showed the foramen on the affected side to be two to four times the normal caliber (Fig. 1, *a* and *b*). In the 2 remaining patients, in whom the optic foramina were normal, the gross tumors were entirely intra-orbital.

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* Presented at the Annual Meeting of the American Roentgen Ray Society, Washington, D. C., September 30–October 3, 1958.
† The Mayo Foundation, Rochester, Minnesota, is a part of the Graduate School of the University of Minnesota.

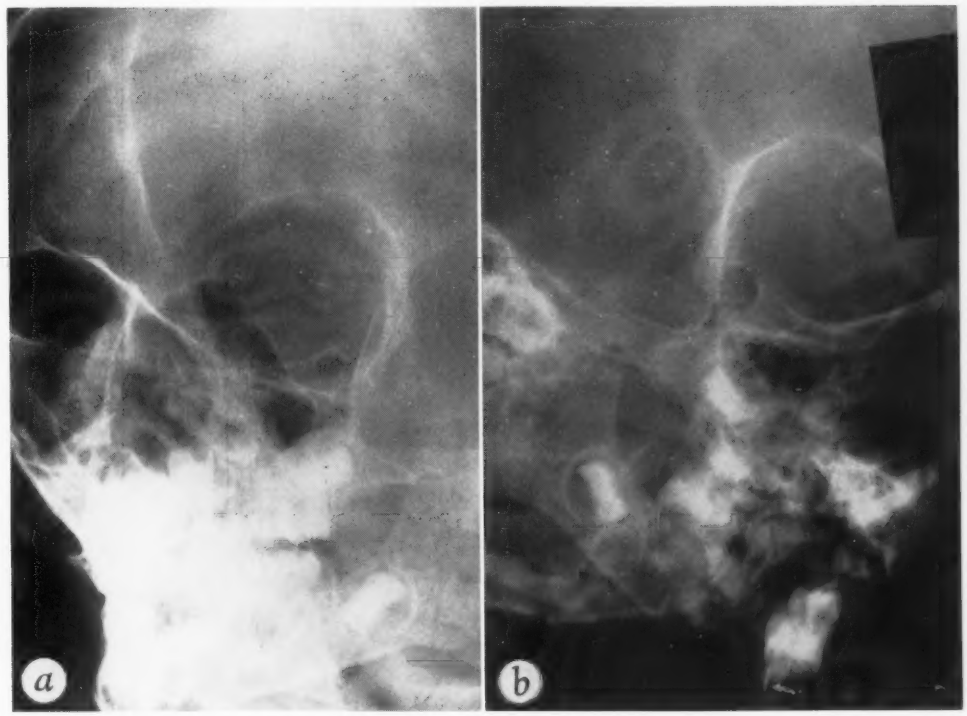


Fig. 1. Optic canal views in a two year old boy with glioma of the left optic nerve. (*a*) Normal right optic canal. (*b*) Left optic canal about four times the caliber of that on the right.

stereoscopic lateral roentgenograms of the heads of 12 of these 13 patients were interpreted as showing no abnormality (Fig. 2). The roentgenograms of 2 patients were not available for restudy. A two year old girl who also had neurofibromatosis exhibited changes similar to those to be described under tumors of the optic nerve and chiasm. Both optic foramina were considerably larger than normal; they were round in cross section, and the margins were not decalcified. A large tumor was removed from the right orbit and optic foramen. The left optic nerve and the optic chiasm appeared to be normal at this time, but the surgeon's impression was that the patient might have an astrocytoma of the left optic nerve. Clinically, a choked disk was present on the left. Follow-up ophthalmologic examination one year later revealed elevation of the left optic disk of 1 diopter and chronic papilledema. Perhaps more time should pass before one may be certain that this patient does not have a tumor of the left optic nerve.

### TUMORS OF THE OPTIC NERVE AND CHIASM

The remaining 35 patients in this series had gross involvement of the optic chiasm and of one or more of the emerging optic nerves. In 4 of these patients, the routine roentgenograms of the head had been interpreted as being normal; the roentgenograms in 3 of these were not available for study.

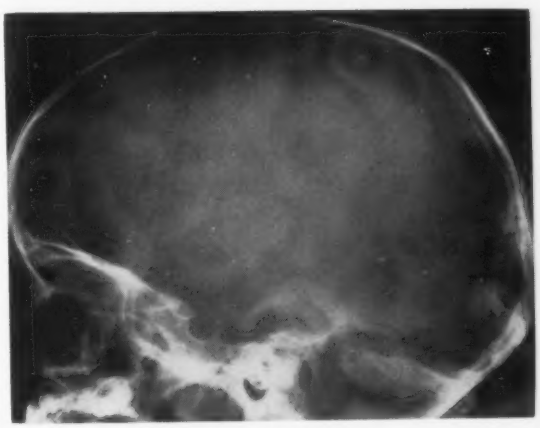

Fig. 2. Same case as in Figure 1. Lateral view shows normal calvarium.

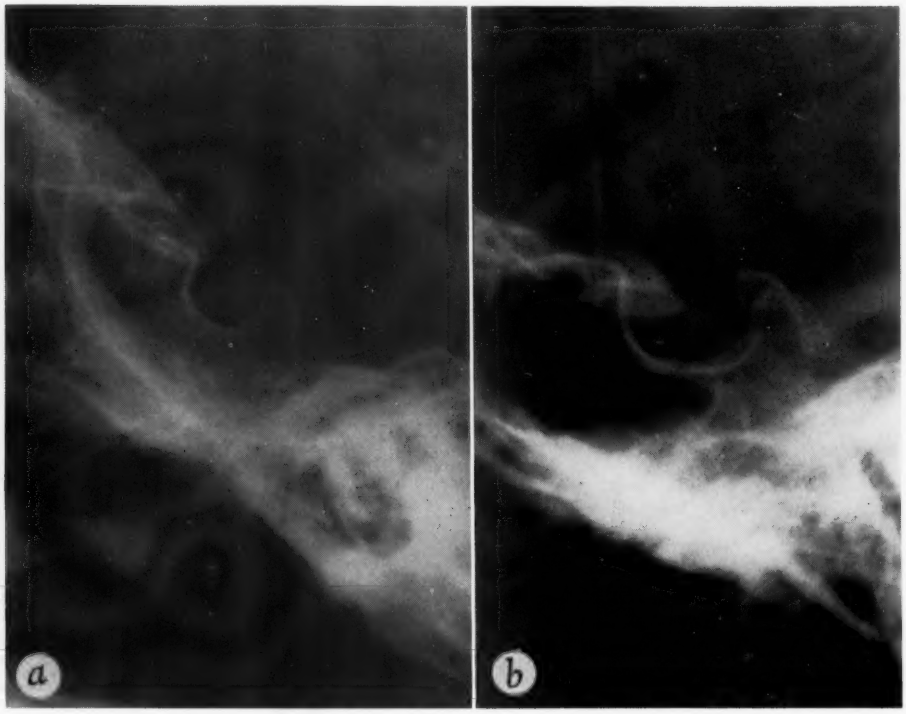


FIG. 3. (*a*) Normal infantile type of sphenoid. (*b*) Normal fully developed adult type of sphenoid.

Most of the changes observed in the other patients involved portions of the sphenoid bone extending from the planum sphenoidale posteriorly to the dorsum sellae.

In the newborn, the floor of the cranium is relatively shorter than it is in older children and adults. A change occurs during growth in the relative size of the facial and sphenoid bones with respect to those of the calvarium.[22] Of importance in this regard is the relatively lower position of the pituitary fossa with respect to the level of the planum sphenoidale and floor of the anterior fossa in the newborn as compared to that usually found in older people (Fig. 3, *a* and *b*). This was alluded to by Goalwin[8] in his discourse concerning the change in the axis of the optic canal that takes place during growth. The effect in the newborn is that of an elongated, sloping or irregular region between the limbus of the planum sphenoidale and the tuberculum sellae proper. This is the region of the chiasmatic sulcus, of which more will be said.

The craniofacial disproportion of the infant results in, or is associated with, a relatively lower position of the optic canal, which allows these foramina to be identified in the usual posteroanterior roentgenogram of the head in patients of this age group. Such a feature is well illustrated in Case 3 of Hertz and Rosendal.[11] This is not ordinarily true later in development unless the usual pattern of growth has been altered.

In 4 of our patients, whose ages ranged from two months to six years, an enlarged optic foramen was seen on the posteroanterior projection of the head (Fig. 4). The chiasm as well as the nerve was involved by the tumor in all these patients.

Martin and Cushing[19] described an appearance sometimes accompanying such tumors that has become known widely as the J-shaped, gourd-shaped or pear-shaped sella. In such cases, pronounced upward concavity of the sphenoid bone is present between the pituitary fossa and the planum sphenoidale, extending usually to one of the optic foramina and involving the anterior clinoid process on that side (Fig. 5). Such an appearance was noted in 8 of these 35 patients. A distinct, although less pro-

nounced, upward concavity of the sphenoid was present in the region of the chiasmatic sulcus in an additional 8 patients. The tuberculum sellae was decalcified in one instance.

Most of these patients had changes involving the region of the sella turcica proper. The pituitary fossa was thought to be enlarged in 12 patients, 3 of whom apparently did not have decalcification of the cortex of the floor or of the dorsum sellae. A total of 13 patients showed some decalcification of cortical bone (Fig. 6). Of these, 4 did not have definite enlargement of the sella turcica. Ten of these patients demonstrated definite shortening or truncation of the dorsum sellae, which usually was not associated with cortical decalcification (Fig. 7). These latter changes are consistent with those that may accompany a suprasellar mass, an intrasellar tumor with suprasellar extension, or extreme enlargement of the third ventricle. They do not appear to be specific for tumor of the optic chiasm.

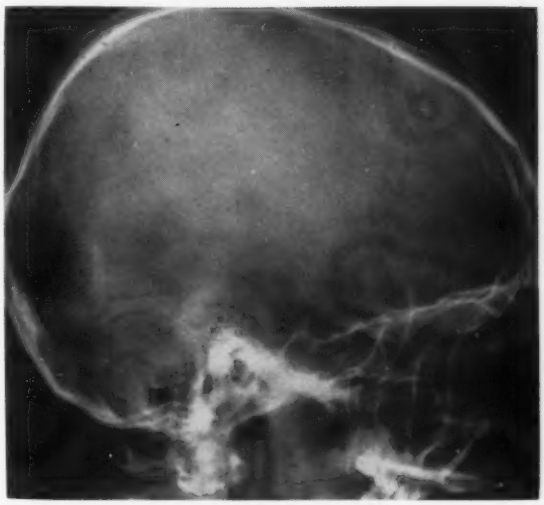

Fig. 5. So-called J sella in a seventeen year old boy with glioma of the chiasm.

Other changes in the cranial vault suggesting the presence of increased intracranial pressure were noted in 7 patients. Five of these showed significant separation of the major cranial sutures, and the other 2 had abnormal and significantly increased prominence of the impressions made by the convolutions on the inner table of the calvarium.

Roentgenograms of the optic foramina were made in 12 of these 35 patients. Def-

Fig. 4. Posteroanterior view in a four year old boy with glioma of the left optic nerve and chiasm. The left anterior clinoid process appears to be projected downward due to enlargement of the optic canal.

Fig. 6. Decalcification of the floor and dorsum of the sella associated with glioma of the chiasm in a twenty-six year old man.

FIG. 7. Truncation of the dorsum sellae. Note also infantile type of sphenoid, with concavity between planum and tuberculum and with decalcification of inferior aspect of anterior clinoid process. Glioma of the chiasm in a nine year old boy.

inite unilateral enlargement of one foramen was found in 4 patients, and bilateral enlargement was noted in 8 (Fig. 8, *a* and *b*).

It is apparent that significant abnormalities occur in routine roentgenograms of the head and optic canal in a large number (90 per cent) of patients who have tumors of the optic chiasm or the optic nerve or both. Unlike the patients reported on by Lindgren and Di Chiro,[17] the present series contained no instances of roentgenologically demonstrable suprasellar calcification.

### SPECIAL EXAMINATIONS

Pneumoencephalography is of value in studying tumors in this region.[2,4,7,10,17] This examination was done in 3 instances in the present series. The gas was injected into the subarachnoid space in the lumbar region of the spinal cord. All 3 patients had obvious obliteration of the cisterns of the optic chiasm and lamina terminalis, as well as a part of the cisterna interpeduncularis. The outline of part of the suprasellar portion of the tumor could be detected in this latter region. Convexity of the floor of each frontal horn of the lateral ventricles was noted in 2 patients, and elevation and posterior displacement of the anterior portion of the third ventricle were found in one. All these patients apparently had slight posterior dislocation of the aqueduct of Sylvius, but this observation needs further evaluation (Fig. 9, *a* and *b*).

Four ventriculographic examinations

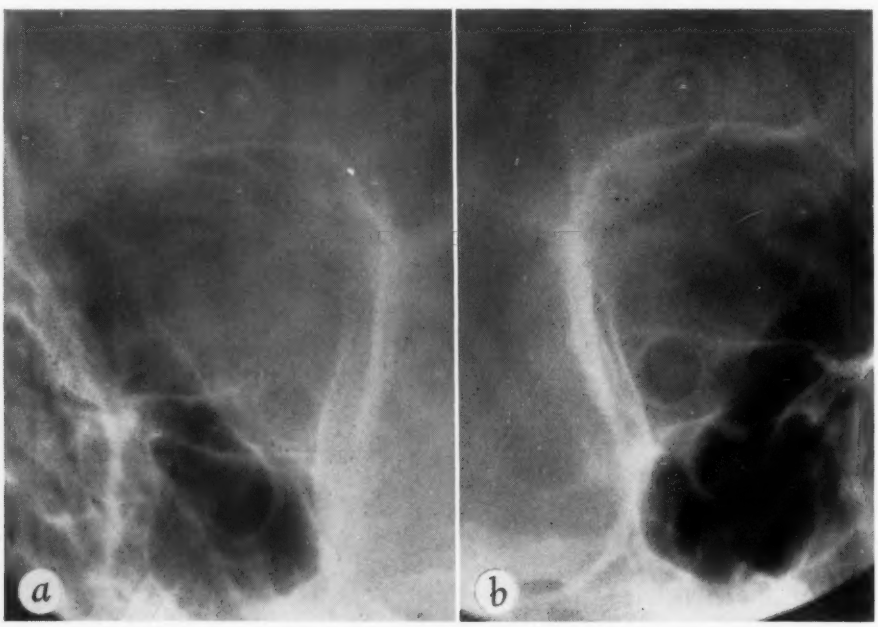


FIG. 8. Glioma of the chiasm in a nine year old boy with great enlargement of both optic canals. (*a*) Right optic foramen. (*b*) Left optic foramen.

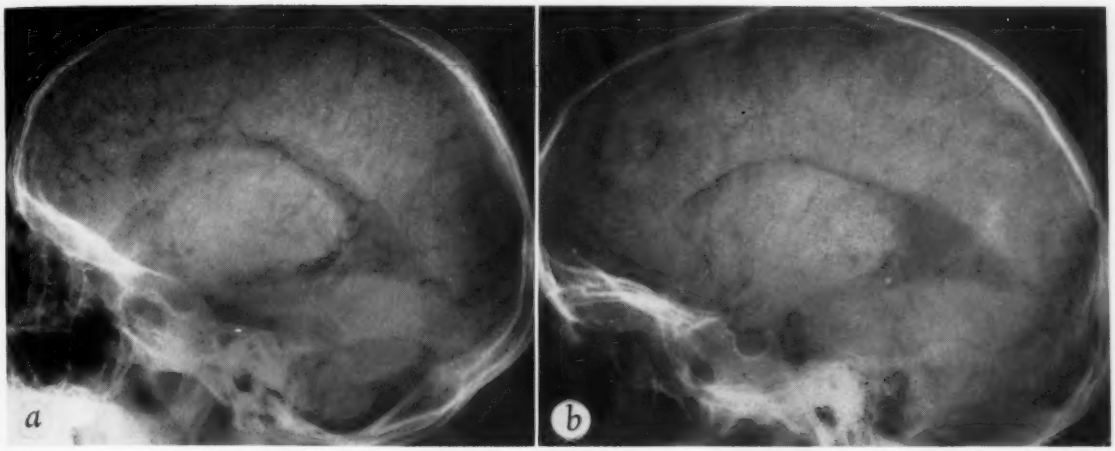


Fig. 9. (*a*) Large glioma of the chiasm in a seven year old girl. Note the changes involving the sphenoid and anterior clinoids, as well as the pneumoencephalographic features described in the text. (*b*) Large glioma of the chiasm in a six year old boy. The tumor is adjacent to the concave portion of the sphenoid between the limbus of the planum and the tuberculum and is partly outlined in the interpeduncular cistern.

were made, 2 of which disclosed changes in the floor of the frontal horns as noted in the pneumoencephalograms. However, all 4 patients had obstruction at the foramen of Monro, with symmetric dilatation of the lateral ventricles; this obstruction was incomplete in one instance. These procedures showed the presence of a suprasellar mass and indicated to some degree the size of the tumor (Fig. 10).

Carotid angiography was undertaken in 2 patients. In one of these, the nerve only was found to be involved at operation. Neither of these 2 examinations gave convincing evidence of dislocation of the major vessels in the region of the sella turcica. A shortened or truncated dorsum sellae was present in the one patient who had involvement of the optic chiasm (Fig. 11, *a* and *b*).

## COMMENT

It appears probable that a large optic foramen occurs in young children as a result of growth of the tumor in the nerve during the time that the sphenoid bone is developing. This would be consistent with the fact that the margins of the canals are not decalcified, as might be expected should a tumor expand fairly rapidly within a fixed bony canal and exert pressure on its walls. This also is suggested by the fact that tumors of this type sometimes are present at birth and, therefore, before ossification of the sphenoid is complete.

Correct evaluation of changes involving the optic foramen depends on meticulous roentgenography. De Kleyn and Stenvers[16] first suggested that the Rhese ethmoid position was useful in the study of these canals, but the detailed studies of Goalwin[9] clarified the need for accurate projection. Because the bony canal has an appreciable length, its margins will not produce a sharp

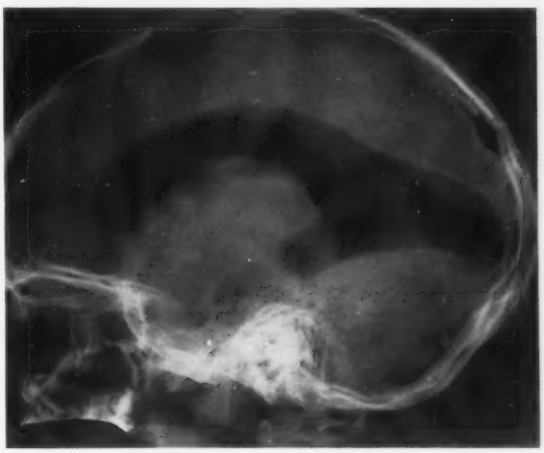

Fig. 10. Large glioma of the chiasm in a five and one-half year old girl. Note the prominent changes in the sphenoid, as well as the ventriculographic features described in the text.

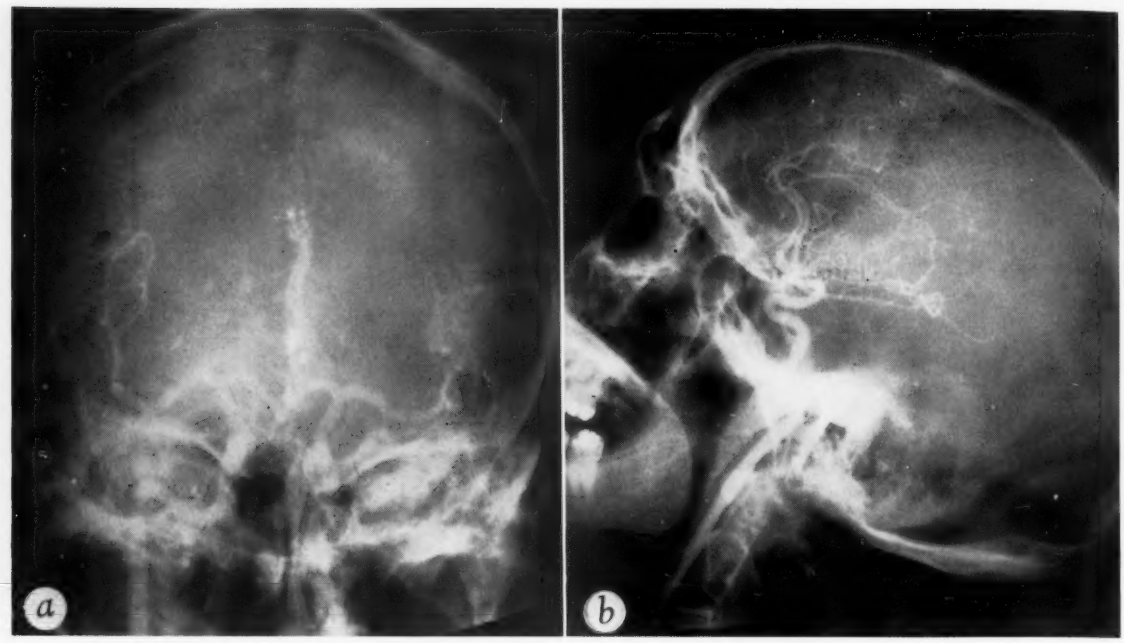


FIG. 11. Large glioma of the chiasm in a thirty-four year old man. The vessels were not displaced but coursed through the tumor. (*a*) Half-axial projection: arterial phase. (*b*) Right lateral projection: arterial phase.

image unless the axis of the central beam coincides with that of the canal. Thus, faulty projection may lead to an erroneous diagnosis of marginal decalcification.

In the present series of cases, those optic canals which were larger than normal were almost uniformly round in cross section. There were no instances of selective decalcification of part of the bony wall, as has been described accompanying extradural carotid aneurysm,[14] pituitary adenoma[18] and other intracranial tumors not primarily involving the optic nerve.[6,13,21] Olivecrona[20] described 2 patients with proved cholesteatomas who had abnormally large optic canals that were rounded in cross section and who also had sphenoidal outlines typical of the J-shaped or gourd-shaped variety. Parts of these tumors apparently surrounded the chiasm and optic nerves and extended through the canals. He observed that cholesteatomas also may be congenital and so it is not surprising that during skeletal growth the sphenoid may adapt itself to the growing tumor in the same manner suggested in the case of optic nerve tumors growing slowly in young children.

It is not easy to assess the significance of some of the changes described in the shape of the sphenoid anterior to the sella turcica. It is particularly difficult to evaluate the significance of the J-, gourd- or pear-shaped sella turcica. Goalwin[8] stated that this configuration is not due to enlargement of the optic canals but rather to deepening and enlargement of the chiasmatic sulcus with or without erosion of the under surfaces of the anterior clinoid process. He indicated that this was proved because he had seen examples of enlarged canals without deformity of the chiasmatic sulcus. Of the 3 patients in this series who had such a sphenoidal configuration and who also had special roentgenograms of the optic foramina, all showed abnormally large canals that were round in cross section and without marginal decalcification. Two other patients had bilateral enlargement of the optic foramina without a J-shaped sella turcica. One of these had a greatly shortened or truncated dorsum sellae, a definite upward concavity of the sphenoid between the pituitary fossa and the planum sphenoidale (the chiasmatic sulcus), and abnormal separation of the cranial sutures. The other pa-

tient had an elongated, slightly concave, upward outline of the region of the chiasmatic sulcus. A pneumoencephalogram in this instance showed the tumor above the sella turcica (Fig. 9*b*).

This view is consistent with the discussion by Ingalls following the paper of Keyes.[15] It may be that the sphenoid in the region of the chiasmatic sulcus and the optic foramen accommodates itself in some cases to a growing mass within the chiasm and emerging nerve. The bone may develop around the mass, thus explaining why it does not show evidence of pressure decalcification in its cortex. This is similar to the molding of the inner table of the calvarium by the growing brain. The time of origin of such tumors with relationship to the growth and development of the sphenoid may be important. Tumors arising later, or growing more rapidly at another time, may not be able to mold the growing sphenoid. However, they may have some effect on its development, since so many of these patients have sphenoids the shape of which in the region of the chiasmatic sulcus appears to be much more similar to that of the infantile type of sphenoid than to that seen in the fully developed and pneumatized adult variety.

One cannot rely on this configuration for the diagnosis of glioma of the optic chiasm, for sphenoids of extremely similar shape are found in a high percentage of patients with mongolism,[25] and such sphenoidal outlines have been observed in patients with neurofibromatosis in whom clinical evidence of a tumor involving the optic nerve was absent.[3] Indeed, the bony dysplasias of the skull, especially those involving the orbit, that sometimes accompany this condition are well known (Fig. 12). This configuration sometimes is seen in the presence of suprasellar cholesteatoma[20] and in otherwise normal persons, as well as in cretinism, dyschondroplasia and rickets. However, all these conditions apparently are related to congenital or developmental abnormalities. Thus, in the present stage of incomplete understanding of the significance of this peculiarly shaped sphenoid, one should

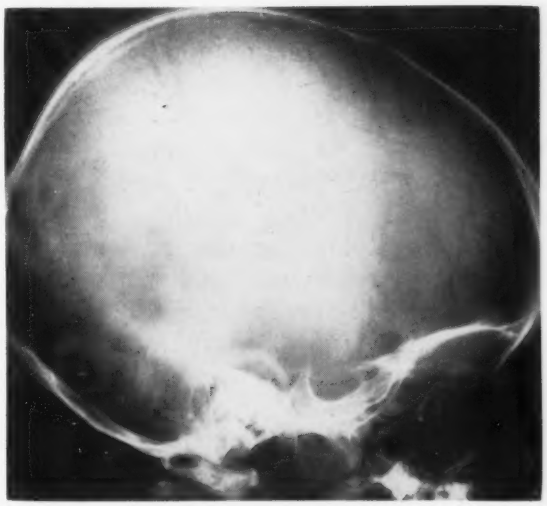

Fig. 12. Similar deformity of the sphenoid in a young child with neurofibromatosis.

not make an unequivocal diagnosis of tumor of the optic chiasm from this appearance alone, although it may be a good diagnostic possibility in the presence of clinical involvement of the optic apparatus. However, enlargement of both optic foramina, together with a J-shaped sella, certainly warrants careful evaluation for possible tumor of the optic nerve and optic chiasm.

The other osseous changes involving the structures about the sella turcica in this series were similar to those accompanying many other suprasellar tumors. Shortening or truncation of the dorsum sellae was noted frequently, but the cortex of the dorsum sellae usually was not decalcified in these cases. The pituitary fossa in some instances was enlarged, and the cortex of its floor was decalcified.

Unlike most other suprasellar tumors that involve or alter the anterior clinoid processes, these structures, when affected, appeared to be decalcified from below rather than from above. As a result, the inferior portions, especially at the base near the optic foramen, appeared to be thinned and decalcified in many cases.

Changes in the vault included separation of the cranial sutures and an abnormal prominence of the convolutional impressions. The intracranial pressure was in-

creased in these cases due to obstruction of the foramen of Monro. The pneumoencephalographic and roentgenologic features were those expected with a suprasellar mass in the region of the chiasm.

It is reasonable to expect cerebral angiography to be of value in delineating these tumors involving the chiasm,[24] although changes were not recognized in the one patient in this series who had both a chiasmal lesion and angiographic studies. However, experience with special diagnostic studies in these lesions is necessarily limited, because the clinical, ophthalmologic and general roentgenologic features are usually sufficient to establish the diagnosis.[5] It may be that more extensive use of special studies might be of assistance in defining the size of the lesions in preparation for treatment.

## SUMMARY

A study was made at the Mayo Clinic of the roentgenograms in 48 proved cases of glioma involving the optic nerve, optic chiasm or both. Routine roentgenograms of the head were normal in 12 of 13 patients in whom one of the optic nerves only was involved. Seven of the 9 patients in this group who had roentgenograms made in the Rhese position had gross enlargement of the optic foramen on the affected side; in the other 2 patients, the tumors were entirely intraorbital.

The remaining 35 patients had gliomas that involved the optic chiasm. In only 4 of these were the roentgenograms considered to be normal. The region of the chiasmatic sulcus showed a large concave depression in 8 patients and a more shallow depression in another 8.

Seven patients of this group had roentgenologic evidence of increased intracranial pressure. There was no instance of roentgenologically demonstrable tumor calcification in the entire series.

Pneumoencephalography was done in 3 cases, in all of which it demonstrated obliteration of the cisterna chiasmatis and lamina terminalis, posterior dislocation of the anterior portion of the third ventricle, and an outline of part of the suprasellar tumor in the cisterna interpeduncularis.

Ventriculography, done in 4 cases, showed severe obstruction of the foramen of Monro in 3 instances and incomplete obstruction in one.

Angiography was done in 2 cases; the results in both were considered to be normal.

## CONCLUSIONS

Gliomas of the optic nerve or chiasm or both are accompanied by roentgenographically demonstrable abnormalities in a large majority of cases.

Tumors limited grossly to the optic nerve usually are associated with a large rounded optic canal without decalcification of its margin.

Almost half of the tumors involving the chiasm are associated with changes involving the inferior aspect of the anterior clinoid processes and that portion of the sphenoid bone between the limbus of the planum and the tuberculum. This is thought to be related to growth of the tumor in the developing infantile calvarium. Although not pathognomonic of such a tumor, these sphenoidal changes, when accompanied by enlargement of one or both optic foramina or in the presence of an appropriate visual disturbance, should suggest the possibility of glioma of the optic chiasm.

Other gliomas of the chiasm frequently present the general and special roentgenographic characteristics of other suprasellar masses.

An estimate of the size and extent of the suprasellar tumor may be obtained by pneumoencephalography and ventriculography.

The Mayo Clinic
Rochester, Minnesota

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# PSEUDOSUBLUXATION OF THE AXIS IN CHILDREN*

*By* GEORGE JACOBSON, M.D., *and* H. HARLAN BLEECKER, M.D.
LOS ANGELES, CALIFORNIA

CHILDREN are frequently seen, following injury to the neck, in whom a lateral roentgenogram of the cervical spine shows an anterior displacement of the axis on C-3. This finding has often been misinterpreted as representing a true traumatic dislocation, with the result that these patients have been subjected to prolonged and unnecessary treatment.[2,4,6]

In reviewing the injuries to the cervical spine seen at the Los Angeles County Hospital from 1950 to 1956, 10 cases involving children between the ages of four and six were found which had been diagnosed and treated as anterior dislocations of the axis. Of these only 4 representative cases are being reported in detail. In spite of an extremely large traumatic service, other injuries to the cervical spine recorded in this age group were most uncommon and did not differ either in type or distribution from those encountered in adults.

## REPORT OF CASES

CASE I. F.C., No. 1230-914. The patient, a six year old Negro female, was admitted to the Los Angeles County Hospital on August 8, 1950. She had fallen from a swing the day before and immediately afterwards complained of pain in her neck. Because the symptoms persisted, the child was brought to the hospital. On physical examination it was noted that she held her head in flexion and that she complained of pain with either active or passive motion of the neck. There were no other positive physical findings.

The roentgenograms of the cervical spine were interpreted as showing an anterior subluxation of the axis on C-3 with considerable angulation at this level (Fig. 1*A*). In forty-eight hours she was asymptomatic and a repeat examination showed that the anterior displacement of C-2 was no longer present. She was placed in a head halter for two weeks and then discharged from the hospital in a Minerva jacket. The patient remained in the jacket for eight months and a roentgen examination at that time (Fig. 1*B*) showed a partial recurrence of the subluxation of C-2. Though she was asymptomatic and there were no positive physical findings, she was re-admitted to the hospital and again placed in head halter traction. A lateral roentgenogram of the cervical spine again showed a disappearance of the subluxation (Fig. 1*C*) and a plaster cast was reapplied. The cast was finally removed and the patient was discharged from care twenty months after the original injury.

*Comment.* While there are several millimeters of anterior displacement of C-2 on C-3 in Figure 1*A*, it should be noted also that the head and neck are sharply flexed. In each of the subsequent examinations, the amount of forward displacement of C-2 varied with the degree of flexion. In Figure 1*B*, while there is no flexion of the neck, the head has been allowed to drop forward and there is a minimal offset of C-2. When the roentgen examination was made with the head and neck in true neutral position as in Figure 1*C*, no displacement or angulation was present.

CASE II. J.E., No. 933-103. The patient, a six year old Caucasian male, was admitted to the Los Angeles County Hospital on July 8, 1952. He had fallen from a tree two days previously, and the following morning awakened complaining of pain in his neck. This pain continued and he was brought to the hospital. Physical examination showed that the head was held in flexion and that the child complained of pain on any attempted active or passive motion of the neck.

Roentgenograms of the cervical spine (Fig. 2*A*) showed slight anterior displacement of C-2 and C-3 with moderate angulation at that level. The following day he was asymptomatic and roentgenograms showed a normal alignment of the vertebrae (Fig. 2*B*). He was placed in a Minerva jacket and discharged from the

* From the Departments of Radiology and Orthopedics, School of Medicine, University of Southern California and the Los Angeles County Hospital.

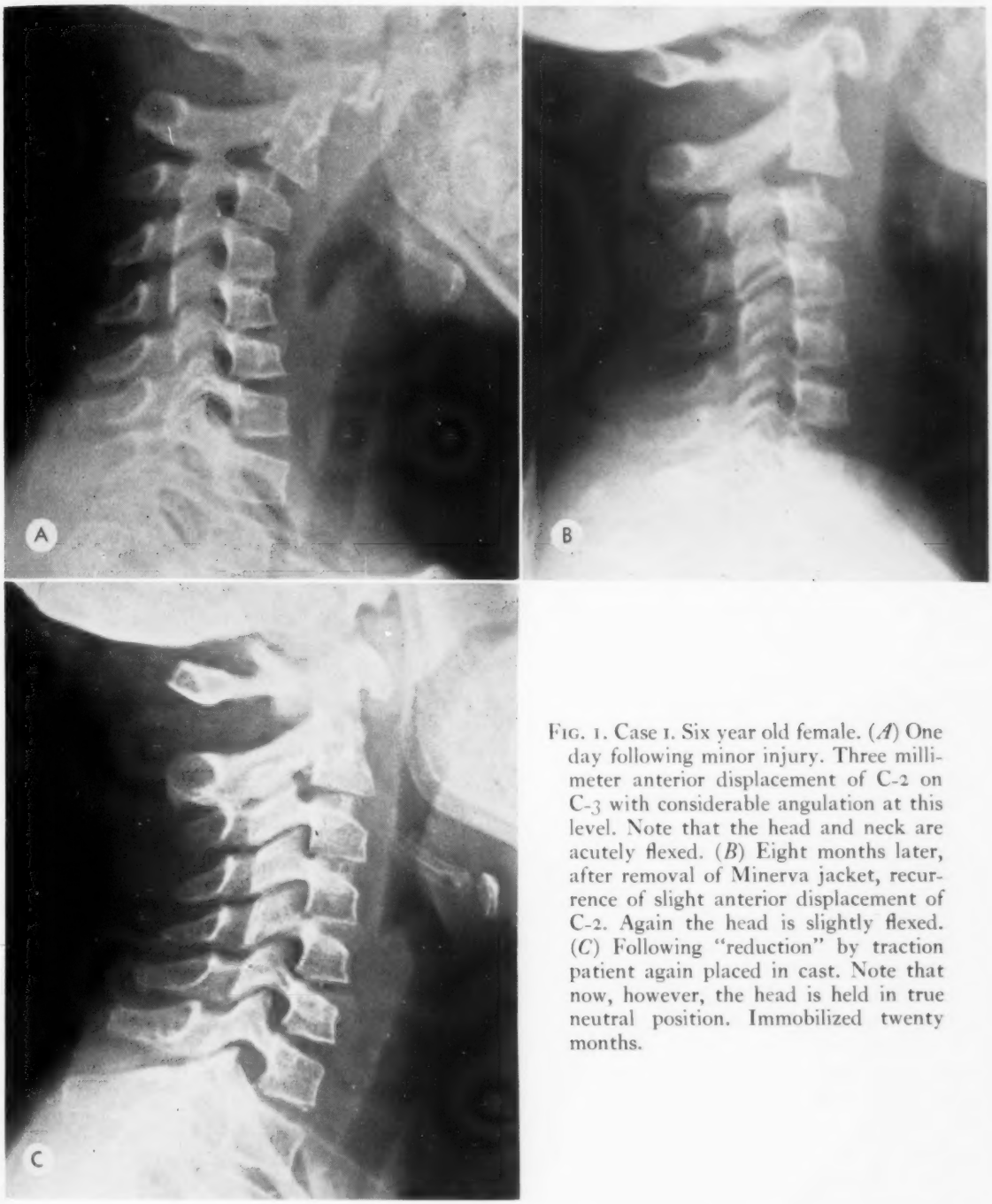


Fig. 1. Case 1. Six year old female. (*A*) One day following minor injury. Three millimeter anterior displacement of C-2 on C-3 with considerable angulation at this level. Note that the head and neck are acutely flexed. (*B*) Eight months later, after removal of Minerva jacket, recurrence of slight anterior displacement of C-2. Again the head is slightly flexed. (*C*) Following "reduction" by traction patient again placed in cast. Note that now, however, the head is held in true neutral position. Immobilized twenty months.

hospital. On September 21, 1952 a roentgenogram taken with the head held in flexion showed a recurrence of the displacement. Immobilization was continued and on re-examination through the cast the vertebral bodies were again in normal alignment. On January 15, 1953 (Fig. 2*C*) an examination made through the collar again showed the displacement. He remained in the collar until May 18, 1953, for a total period of ten months. A roentgenogram made two years later at the age of eight (Fig. 2*D*), with the head slightly flexed, again showed a minimal angulation at the C-2/C-3 level.

*Comment.* The amount of displacement

A
B
C
D

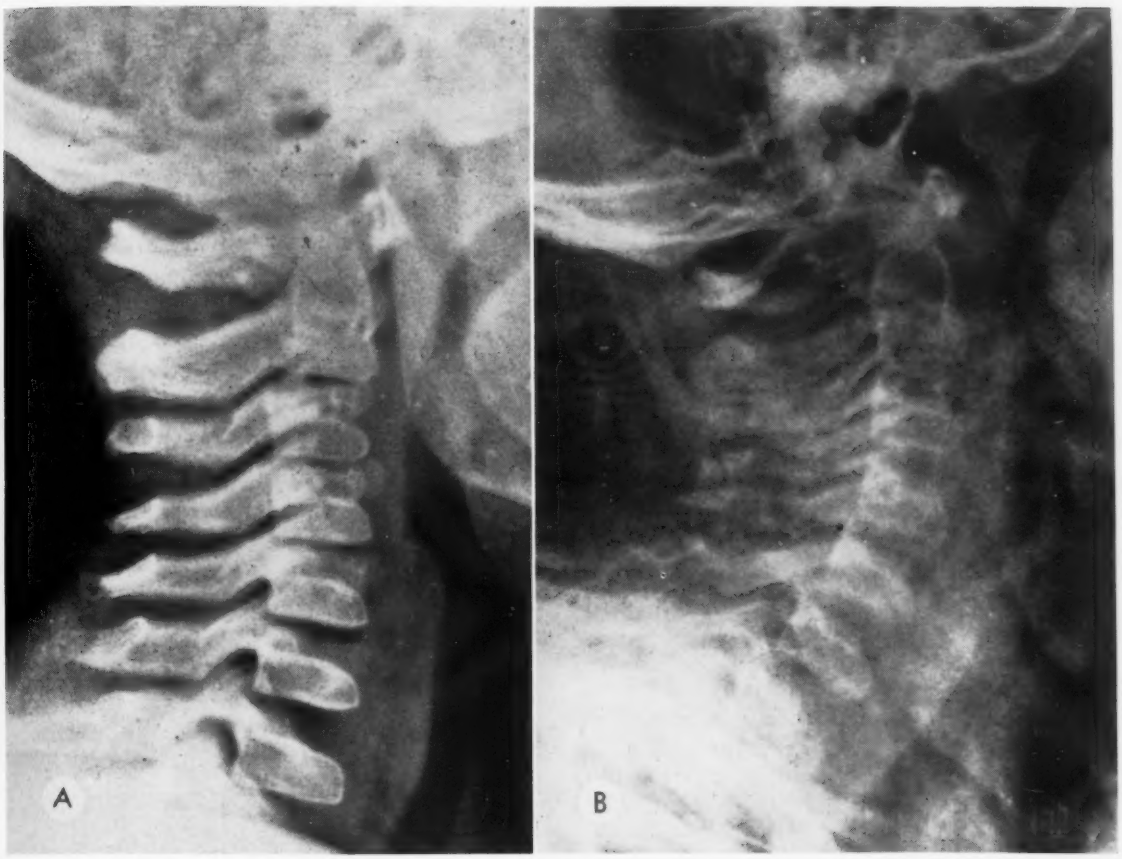


Fig. 3. Case III. Six year old male. (*A*) Several hours after minor injury. Minimal anterior displacement of the axis without angulation. Note that the head is slightly flexed. (*B*) Four days later the head and neck are extended and the vertebral bodies form a smooth concave curve. Immobilized eleven months.

and angulation seen on the original roentgen examination of this patient, while not as striking as in Case I, is that which is more commonly encountered. The numerous roentgen examinations amply demonstrated the effect of flexion of the head and neck on the relationship of C-2 to C-3.

Case III. J.G., No. 1434-539. The patient, a six year old Caucasian male, was admitted to the Los Angeles County Hospital on September 5, 1953, several hours after having fallen from his bicycle. His chief complaint was pain in the neck. On physical examination it was found that he held his head in flexion and that he complained of pain with any motion of the neck.

Roentgenograms of the cervical spine (Fig. 3*A*) revealed a minimal anterior displacement of C-2 on C-3 without any significant angulation. He was placed in head halter traction and became asymptomatic after twenty-four hours. Three days later he was placed in a Minerva jacket and discharged from the hospital. Roentgenograms at that time showed normal alignment of C-2 and C-3 (Fig. 3*B*). He was kept immobilized for eleven months.

*Comment.* This is an example of minimal

Fig. 2. Case II. Six year old male. (*A*) Two days following minor injury. Moderate angulation at C-2/C-3 level with slight anterior displacement of C-2. Note the flexion of the head and neck. (*B*) The following day the head is extended and the "subluxation" has been reduced. (*C*) Six months later an examination through the cast shows a recurrence of the original deformity. The head is again flexed. (*D*) Two years later slight flexion of the head is accompanied by minimal angulation at the C-2/C-3 level. Immobilized ten months.

displacement of the axis without the usual angulation. Note that in Figure 3*A* the head is flexed. When as in Figure 3*B* the head is extended, the anterior displacement of the axis is no longer present.

CASE IV. R.K., No. 1693-558. The patient, a six year old Negro female, was admitted to the Los Angeles County Hospital on September 24, 1956. She had been pushed off a swing several hours prior to entry. On physical examination it was found that her head was held in flexion and that she complained of pain with attempted active or passive motion of the neck.

Roentgenograms of the cervical spine (Fig. 4*A*) showed moderate angulation between C-2 and C-3 without any demonstrable displacement of C-2. After forty-eight hours of traction in a head halter, she became asymptomatic. She was placed in a Minerva jacket and discharged from the hospital. Her neck was immobilized for five months. Roentgenograms taken while she was in the Minerva jacket showed normal alignment of C-2 and 3-C (Fig. 4B). Roentgenograms in flexion and extension (Fig. 4, *C* and *D*) after removal of the jacket showed not only angulation at the C-2/C-3 level, but also forward displacement of C-2.

*Comment.* This case illustrates angulation of the C-2/C-3 level without displacement. Note that following immobilization (Fig. 4*C*) a greater degree of flexion of the head than in the original examination (Fig. 4*A*) has produced anterior displacement of C-2 as well as angulation at this level.

## DISCUSSION

The clinical picture presented by these patients, including the 6 not described in detail, was quite similar: a minor injury followed in a short time by pain on motion of the head and neck, varying degrees of torticollis, and roentgenograms of the upper cervical spine which were interpreted as abnormal. All became asymptomatic after twenty-four to forty-eight hours of immobilization.

The lateral roentgenograms of the upper cervical spine showed either anterior displacement of C-2 on C-3 and/or angulation at the same level. These findings were considered to be pathologic in view of the fact that they are not encountered in the normal adult and that at that time we did not have a clear concept of the normal range of motion in children.

## NORMAL FLEXION AND EXTENSION OF THE CERVICAL SPINE

At all ages, when the head and neck are in the neutral position, the posterior border of the cervical spine usually forms a smooth concave curve. With flexion the concavity is progressively reduced, the posterior border becomes more or less straight, and with acute flexion may even become convex.

During infancy and childhood up to approximately the age of ten, flexion and extension are centered at the second and third cervical vertebrae. Figures 5, 6 and 7 are representative of the changes which occur with flexion and extension in this age group. In each example it can be seen that with flexion, angulation takes place at the C-2 and C-3 level and that the spine below this presents different degrees of concavity and convexity. In addition, the relatively horizontal plane of the articular facets and the apparent laxity of the intervertebral ligaments permit a gliding anteroposterior motion between these two vertebrae. This may vary from little or no forward displacement, as in Figure 5*A*, to several millimeters, as in Figure 7*A*. As much as 5 mm. has been demonstrated by us in a normal child. Posterior gliding of C-2 on C-3 may be produced by hyperextension, as in Figure 5*C*, though this is less frequent.

At the age of ten or thereabouts, in both males and females, the apex of the flexion curve shifts downward to the C-4/C-5 or C-5/C-6 level (Fig. 8), where it remains. We have not observed forward gliding of C-2 on C-3 in normal older individuals, though a step-like offset may occur at lower levels.

A critical evaluation of the relationships of the second to the third cervical vertebrae in the children treated for subluxation makes it quite apparent that they are entirely within the range of normal motion

Fig. 4. Case IV. Six year old female. (*A*) Several hours after injury. Moderate angulation at the C-2/C-3 level without displacement of C-2. The head is moderately flexed. (*B*) Two months later with the head and neck extended, C-2 and C-3 form a smooth curve. (*C* and *D*) Five months later roentgenograms made in flexion and extension show not only angulation at the C-2/C-3 level, but also anterior displacement of C-2. Immobilized five months.

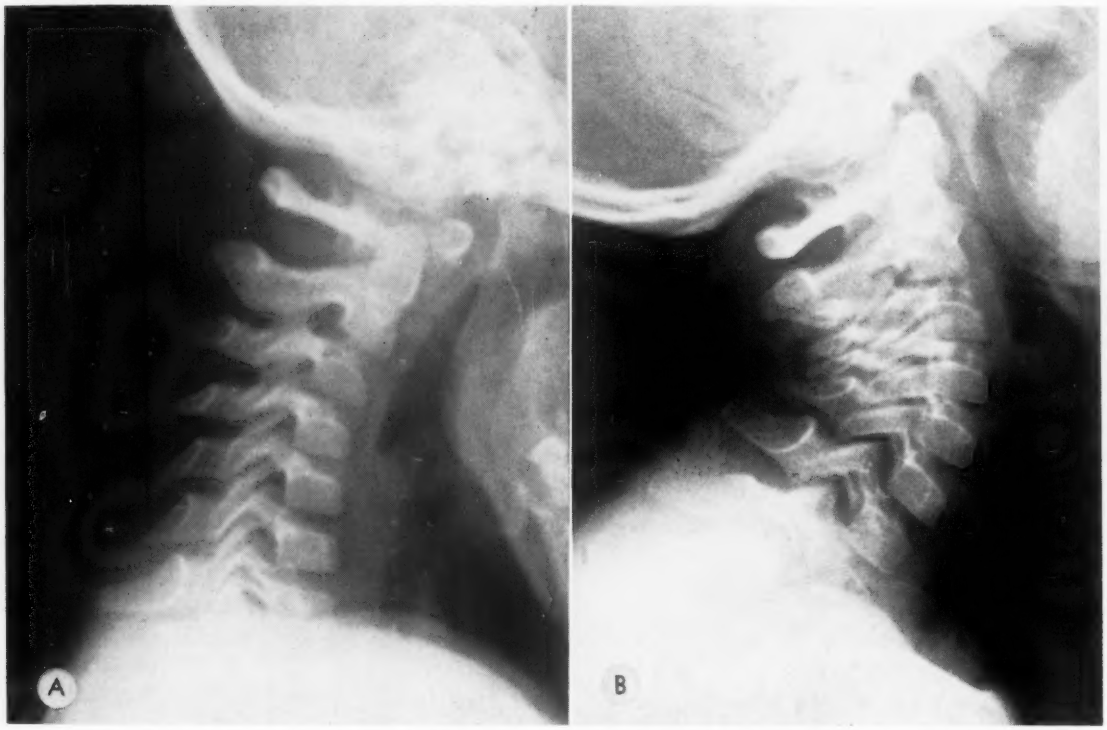


FIG. 5. Normal three year old male. (*A*) Both the head and neck are flexed. The atlas is displaced anteriorly 3 mm. The apex of curvature is at the C-2/C-3 level. (*B*) With extension the anterior displacement of the atlas disappears and the posterior border of the vertebrae forms a smooth concave curve.

FIG. 6. Normal four year old female. (*A*) Only the head is flexed. The atlas is displaced slightly forward. The spine is almost "straight" and there is minimal angulation at the C-2/C-3 level. (*B*) With extension the anterior displacement of C-2 disappears and the spine is smoothly concave.

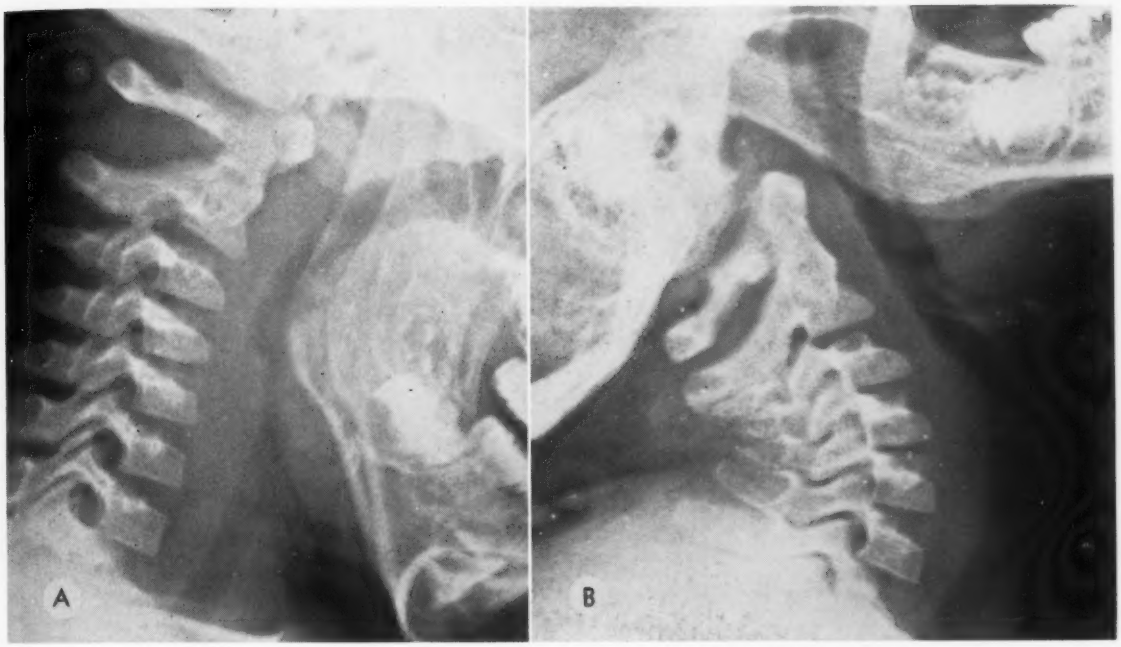


Fig. 7. Normal seven year old male. (*A*) Anterior displacement of the atlas does not always occur even with sharp flexion of both the head and neck. Only moderate angulation at the C-2/C-3 level could be produced in this child. (*B*) Hyperextension. There is minimal posterior displacement of C-2 on C-3.

and can be reproduced easily in normal individuals. Others have commented similarly on the mobility of the axis in children. Bailey[1] in 1952 published an excellent study of the normal cervical spine in infants and children, and noted that during flexion the second cervical vertebra might be displaced anteriorly as much as 2 or 3 mm. In the same year, Townsend and Rowe[7] reported 4 cases of children with forward subluxation of C-2 who were unable to extend their necks to the neutral position following minor or unknown trauma experienced during an upper respiratory infection. The clinical status of each patient improved under treatment, but the roentgen changes failed to show comparable improvement or at times reverted to pretreatment levels without the reappearance of symptoms. As a result, they examined a group of normal children and also noted that the greatest mobility occurred between C-2 and C-3. They remarked that when the head is flexed forward, displacement is to be expected and in their patients was probably accentuated by muscle spasm. Even so, Donaldson[2] in 1956, though aware of the above two papers, described 37 cases of torticollis in patients varying in age from two to thirty-seven years, 75 per cent of whom had an anterior subluxation of C-2. He noted that the history in most of his patients was that of minimal trauma. He also commented that reduction might occur quite easily and quickly, and therefore the roentgenographic examination should be carried out before treatment is instituted. The ages of the patients with subluxation are not given and it is not clear how the diagnosis was established in each case. He advised immobilization for six weeks after reduction by traction. More recently Pendergrass, Schaeffer, and Hodes,[5] and Dunlap, Morris, and Thompson[3] have also pointed out that such cases do not represent true subluxations of the axis.

It should be noted that in each instance in which the axis is displaced anteriorly, or there is an acute angulation at the C-2/C-3 level, either the head or neck is flexed. In general, the greater the degree of flexion, the more marked is the displacement or angulation. *However, only minimal flexion of the head may be sufficient to produce an*

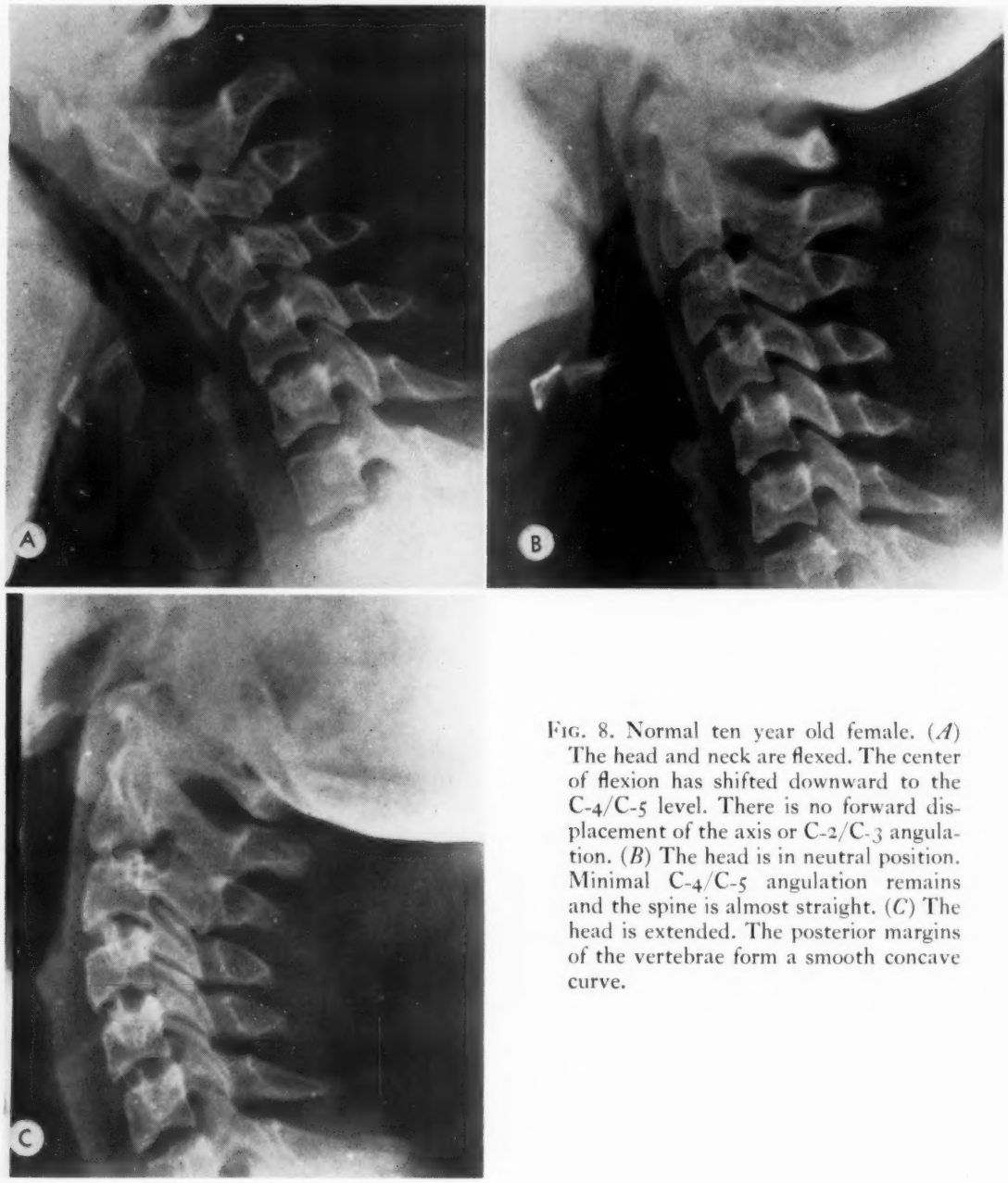


FIG. 8. Normal ten year old female. (*A*) The head and neck are flexed. The center of flexion has shifted downward to the C-4/C-5 level. There is no forward displacement of the axis or C-2/C-3 angulation. (*B*) The head is in neutral position. Minimal C-4/C-5 angulation remains and the spine is almost straight. (*C*) The head is extended. The posterior margins of the vertebrae form a smooth concave curve.

*apparent deformity*. Treatment was frequently prolonged because the head was allowed to fall forward slightly during the examination (Fig. 1*B* and 2*C*). Careful positioning of the head and neck is of paramount importance if even the simulation of a subluxation is to be avoided.

## SUMMARY

1. Ten cases of pseudosubluxation of the axis in children were reviewed and 4 are reported. Fractures or dislocations of the cer-

vical spine are rather unusual in children, and when they do occur do not differ significantly from those seen in adults.

2. Normal flexion and extension of the cervical spine are described and it is pointed out that anterior displacement of the axis and acute angulation at the C-2/C-3 level are normal for individuals up to the age of ten.

3. Children with injuries to the neck and with only the above mentioned roentgenographic findings should be treated symptomatically for whatever soft tissue injury may be present and not for subluxation of the axis.

George Jacobson, M.D.
Department of Radiology
University of Southern California
School of Medicine
1200 North State Street
Los Angeles 33, California

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# VERTEBRA PLANA IN CHILDREN PRODUCED BY XANTHOMATOUS DISEASE

*By* JOSEPH D. CALHOUN, M.D., *and* S. B. THOMPSON, M.D.
LITTLE ROCK, ARKANSAS

## HISTORICAL REVIEW

IN 1925 Calvé[3] published a report of 2 cases of young children who had vertebral body lesions which he thought clinically resembled tuberculous spondylitis but who made a recovery without the usual marked kyphosis, abscess formation, and draining sinuses. Since the roentgenographic appearance of the lesions so closely resembled osteochondritis in other areas of the body, he concluded that osteochondritis was the underlying pathology in these cases. Buchman[2] later suggested the term "vertebra plana." Now, apparently by common usage, Calvé's disease and vertebra plana are more or less synonymous terms employed to denote a flattened vertebral body produced by an underlying osteochondritis. An extensive, but not necessarily exhaustive, search of the literature by us has not uncovered a single instance of vertebra plana proved by microscopic examination to be due to an osteochondritis.

Numerous cases of vertebra plana have been reported in which conditions other than osteochondritis have produced the deformity. Brailsford[1] mentions aseptic necrosis, xanthomatosis, trauma, Gaucher's disease, and Hodgkin's disease as conditions capable of producing vertebra plana. Osteoporosis, metabolic disturbances, malignant tumors, hemangiomas,[8] von Recklinghausen's osteitis fibrosa, and fracture secondary to tetanus[6] have been cited. One can but be impressed by the paucity of histologic proof from biopsy or necropsy material. It was while this communication was in the early phase of preparation that Compere *et al.*[4] published their report of 4 cases of vertebra plana due to eosinophilic granuloma in which 2 had biopsy material taken from the involved vertebral bodies.

The lack of published reports of cases bearing histologic proof stimulated us to report our experience of 4 cases of vertebra plana with histologic diagnoses of eosinophilic granuloma and Hand-Schüller-Christian's disease.

## CASE MATERIAL

Since 1949 we have encountered 4 cases of children having vertebral body lesions which seemed to fit the criteria for "vertebra plana" as described by Calvé. Case 1 had a solitary lesion in a vertebral body, accompanied by severe neurologic changes. A second case had what was thought to be a solitary lesion but "routine" biopsy of the vertebrae above and below revealed unsuspected involvement of the subjacent body. The other 2 cases presented multiple lesions in the skeleton.

## PATHOLOGY

Xanthomatosis is a term that has been suggested to include the three diseases commonly known as eosinophilic granuloma, Hand-Schüller-Christian's disease, and Letterer-Siwe's disease. The literature is clouded with confusion, particularly with reference to terminology in these conditions, and it is not within the scope of this paper to enter this controversy. Perhaps the three diseases are merely "different clinico-anatomic expressions of the same basic disorder" as suggested by Jaffe and Lichtenstein,[7] and eosinophilic granuloma merely represents the chronic phase, Hand-Schüller-Christian's disease the intermediate phase, and Letterer-Siwe's disease the acute phase as suggested by Goodhill.[5] Even though the etiology is uncertain, radiologists and orthopedists are becoming more aware of these diseases, particularly eosinophilic granuloma, in dealing with certain lytic bone lesions.

### CLINICAL FINDINGS

Most of the cases reported are in children from two to ten years of age. Frequently, the history reveals a rather trivial fall with rather insidious development of mild back pain and malaise. The physical findings are usually minimal with only mild tenderness, muscle spasm, and gibbus deformity. There is little to be gained from laboratory procedures. The vertebral lesion may even be discovered incidentally, with the clinical findings pointing to other areas of involvement as in our Case IV.

We are reporting one of our cases (Case I) in detail because of the severe neurologic changes that were present and the marked improvement that followed biopsy and roentgen irradiation. The other 3 cases are reported in abbreviated form.

### ROENTGEN FINDINGS

There have been many detailed descriptions of the roentgen findings of vertebra plana. Some of the more constantly recurring are: (1) Involvement of a single vertebra with the body reduced to a wafer thin disk; (2) preservation of (or slight widening of) the intervertebral cartilage spaces immediately above and below the lesion; (3) slight increase in the anteroposterior diameter of the involved body; (4) a mild kyphosis at the involved level; (5) a relative sparing of the posterior elements; and (6) a period of regeneration which lasts for many months in which the vertical height is nearly or completely restored.

These findings have been more or less true in our 4 cases, except that we have not noted an appreciable degree of restoration of the normal vertical height.

We have been impressed by a gentle, fusiform swelling of the paraspinous soft tissue shadows, extending a vertebra or so above and below the lesion. This shadow has been striking in all but 1 of our 4 cases, and the exception was the case of a lumbar lesion where overlying intestinal gas shadows and psoas shadows might have obscured the swelling. We believe the shadow to be distinctive from the typical paraspinous soft tissue abscess seen in tuberculous spondylitis.

### REPORT OF CASES

CASE I. A.E. About December, 1952 this fourteen year old white male noted that his walking was impaired and there were vague chest pains. He developed stumbling and an unsteady gait which progressed until May, 1953 when he first consulted a physician.

*Physical Examination.* The back examination revealed some prominence of the midthoracic region. No masses were palpated. The patient was barely able to flex the hips and knees because of muscle weakness. The neurologic examination of the back and lower extremities showed a spastic paraplegia with hyperactive deep tendon reflexes, patella and ankle clonus, positive bilateral Babinski sign, hypalgesia and hyperesthesia at D-6 and D-12 levels, and preservation of toe position. There was no fecal or urinary incontinence. The Queckenstedt test showed a complete block.

*Roentgen Findings.* The dorsal spine examination (Fig. 1, *A* and *B*) showed the vertebra plana lesion in the sixth dorsal vertebra. A granulomatous process was considered but it was felt that tuberculous spondylitis could not be ruled out. A skeletal survey revealed no additional lesions.

*Laboratory Findings.* Extensive hematologic, blood chemical, and urine studies revealed no significant abnormal findings.

*Operation.* On June 26, 1953 a needle aspiration (S. B. T.) of the D-6 area was done with thick blood being obtained. Smears were negative for all organisms. On July 14, 1953 a costotransversectomy was performed. The pleura was inadvertently entered and so the procedure was abandoned to allow the pleural rent to seal. On July 29, 1953 the same approach was used. No abscess was found but the body of D-6 was extremely necrotic and the posterior elements friable. Appropriate specimens were taken for microscopic analysis.

*Pathologic Report.* Eosinophilic granuloma of bone (Dr. M. J. Kilbury).

*Course.* The recovery was uneventful except that multiple thoracenteses were necessary to control pneumohemothorax. The boy's spastic paraplegia remained unchanged after the two surgical procedures. Three weeks later he was started on a course of deep roentgen therapy to the D-6 area. A total dose of 1,000 r in air was

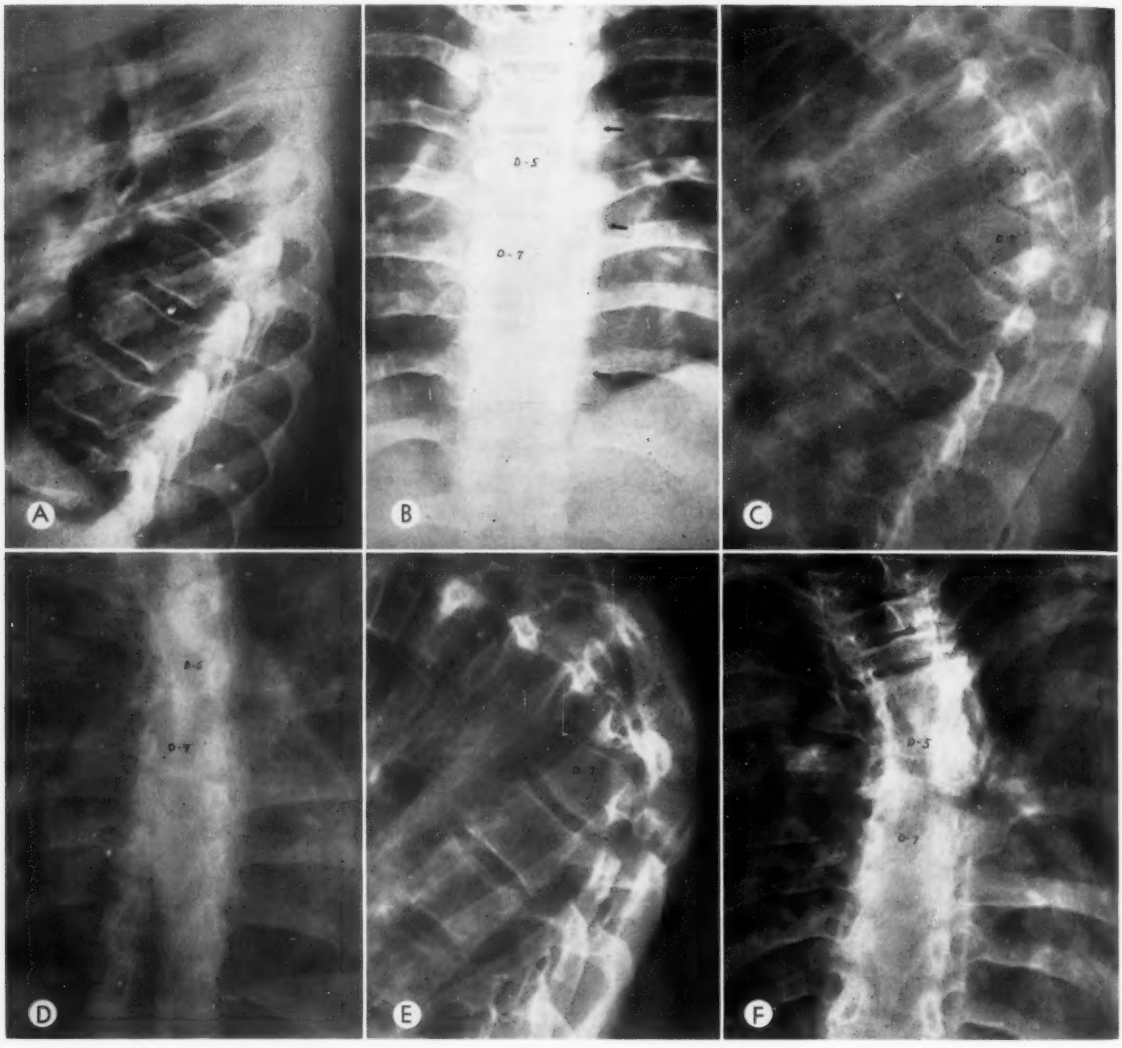


FIG. 1. Case 1. (*A*) Lateral view showing destruction and collapse of the sixth dorsal vertebra and preservation of the adjoining intervertebral spaces. The posterior elements are more involved than in any of the other cases. (*B*) Anteroposterior view showing the gentle fusiform paraspinous bulging. (*C*) Lateral view (eighteen months after surgery and irradiation) demonstrating the increased kyphos (about 90°). (*D*) Anteroposterior view (eighteen months after surgery and irradiation) demonstrating mild scoliosis. (*E*) Lateral view (three years after surgery and irradiation) demonstrating further increased kyphos (about 120°). (*F*) Anteroposterior view (three years after surgery and irradiation) demonstrating slight increased scoliosis.

delivered in five consecutive treatment days (half value layer 1.25 mm. Cu).

A meticulous neurologic examination and muscle evaluation were then done to establish a baseline for future comparison. They did not differ appreciably from the original.

An intensive rehabilitation course was started consisting of various muscle strengthening exercises. This was followed by wheel chair privileges and later resumption of erect walking aided by crutches and bilateral, long leg, double upright braces. By September 25, 1953, the quadriceps had reached a fair or better strength. By October 2, 1953, he was walking with braces and crutches. Within the next week he was able to discard the crutches. The neurologic changes showed gradual clearing. On October 10, 1953, the boy was discharged but there persisted some knee clonus and some hypalgesia in the left lower extremity. A clinic visit on November 3, 1953 showed his general condition greatly improved with a twenty pound weight

gain. He had discarded the braces. A clinic visit on February 3, 1954 revealed that he had resumed school without braces.

The progressively increasing deformity over a three year period after surgery and irradiation (Fig. 1, *C*, *D*, *E*, and *F*) with simultaneous recovery of the neurologic deficit and other clinical symptoms confirms the feeling that the compression of the spinal cord resulted from the granulomatous lesion and not from the skeletal deformity.

One wonders if this patient might have been better off if his spine had been fused sometime in the first year or so after irradiation.

Case II. R. D. B. This seven year old white male sustained a fall from his porch, landing on his back on August 1, 1953. He was asymptomatic for one week when back pain began. His family physician treated him for a week or so for an apparent mild kidney infection. His father had had tuberculosis in 1939 but this was considered to be arrested.

*Physical Examination.* This was negative except for the back where there was a point tenderness over D-10 and D-11 spinous processes, limitation of back motion, and a prominent D-10 spinous process.

*Roentgen Findings.* The dorsal spine examination (Fig. 2, *A* and *B*) showed a vertebra plana lesion of the tenth dorsal vertebra. The lesion was thought to be due to either a benign granulomatous process, trauma or tuberculosis. A bone survey was negative for other lesions.

*Laboratory Findings.* There were no significant abnormal findings.

*Course.* The patient rapidly became asymptomatic on bed rest and was discharged to a convalescent center and later to out-patient care to be followed in clinics (Fig. 2*C*). Although he remained asymptomatic it was thought that a biopsy was advisable because of the family history of tuberculosis. This was done on July 28, 1954 (Drs. J. M. Hundley and F. J. Gray). At surgery it was thought that there might be involvement of D-11 so material was taken from the ninth, tenth and eleventh dorsal vertebral bodies.

*Pathologic Report.* Eosinophilic granuloma of the D-10 and D-11 vertebrae (Dr. W. S. Orr).

*Course.* Recovery was uneventful. He received deep roentgen therapy (half value layer 2.0 mm. Cu) shortly afterward. A dose of 1,300 r in air was given in one week's time to the tenth dorsal area. He is still being followed in clinics and has remained asymptomatic.

*Comment.* Roentgen examination as late as one month before biopsy (Fig. 2 *D*) failed to reveal evidence of destruction of the subjacent eleventh dorsal vertebra. It is conjecturable whether the bony substance was involved or whether the positive material was actually paraspinous extension from the tenth vertebra.

Case III. D. H. This three year old white male child was first seen on July 27, 1953 because his parents thought he had a "weak or stiff" back since he would not bend his back, even while playing. He had a trivial fall two weeks previously.

*Physical Examination.* This was negative except for moderate lower abdominal tenderness and moderate stiffness and weakness of the back muscles.

*Roentgen Findings.* The dorsal spine examination (Fig. 3, *A* and *B*) revealed a vertebra plana lesion of the ninth dorsal body. The skull examination revealed an irregular lytic lesion in the right frontotemporal region as well as a midline frontal lesion (Fig. 3*C*). A complete skeletal survey and chest roentgenogram disclosed no other lesions.

*Laboratory Findings.* There were no significant abnormal findings.

*Operation.* A biopsy was done (S. B. T.) on the frontotemporal lesion.

*Pathologic Report.* Hand-Schüller-Christian's disease (Dr. W. S. Orr).

*Course.* Deep roentgen therapy (half value layer 1.25 mm. Cu) was given to the dorsal lesion and both skull lesions. A dose of 700 r in air was administered to each field in two weeks time. He was then discharged, symptom free, to wear a Taylor back brace which he discarded one year later. He has remained asymptomatic since but has a very slight kyphosis.

Case IV. C. J. This twenty-two month old colored male child was admitted on August 12, 1949 because of a scalp swelling above and behind the right ear for a period of one month and a sore left elbow of five days' duration. Previously the scalp lesion had become fluctuant and drained a dark red material for several days.

*Physical Examination.* There was a fluctuant scalp swelling which was not painful nor feverish. His left arm hung at his side and he cried and fought back attempts of manipulation.

*Roentgen Findings.* A skeletal survey revealed a vertebra plana lesion of the third lum-

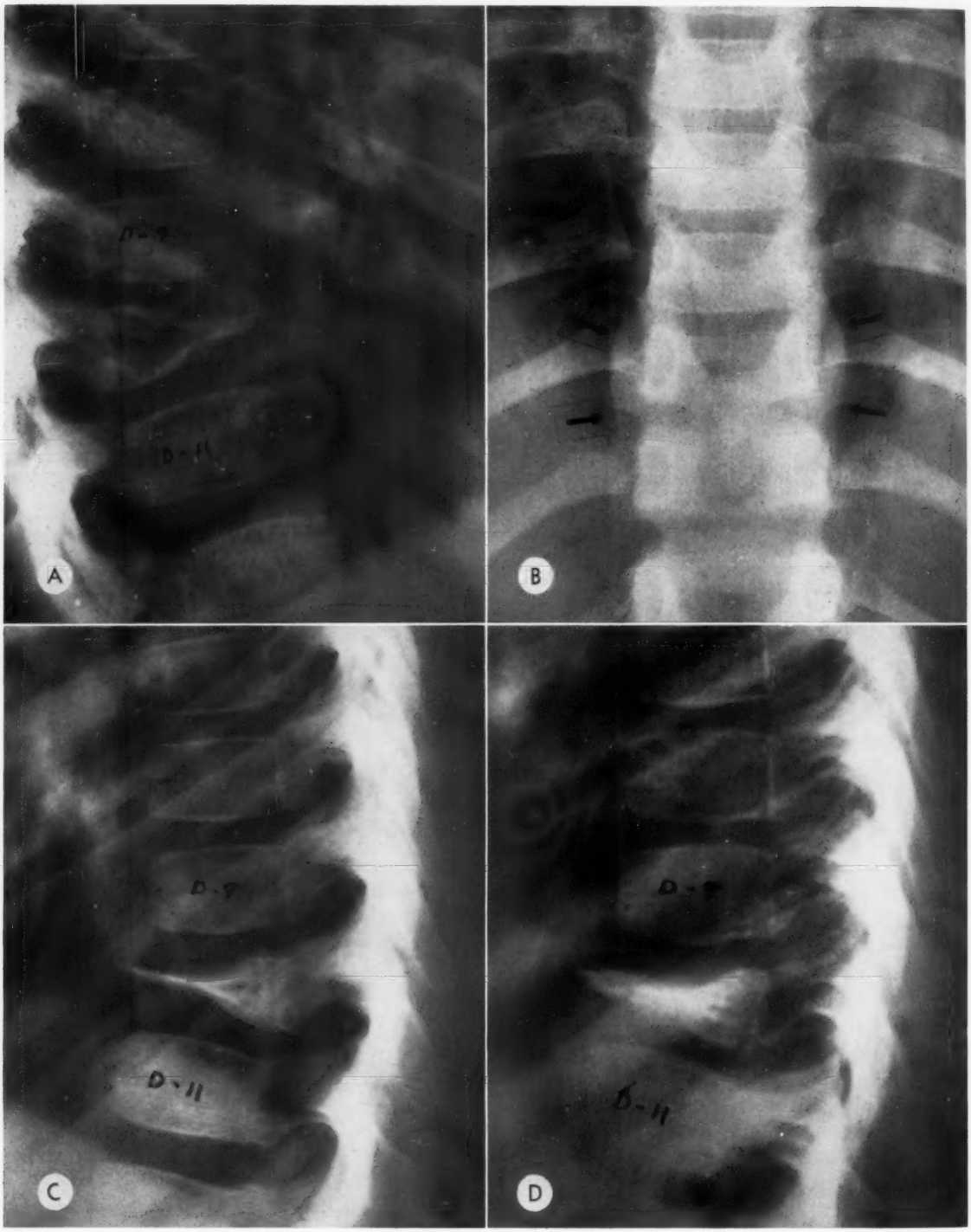


FIG. 2. Case II. (*A*) Lateral view showing collapse of the tenth dorsal vertebra and preservation of adjoining intervertebral spaces and posterior elements. (Courtesy University of Arkansas School of Medicine.) (*B*) Anteroposterior view showing the gentle fusiform paraspinous bulging. (Courtesy University of Arkansas School of Medicine.) (*C*) Lateral view two months later than *A* showing slight alteration. Note more complete collapse of the body and the increase in the anteroposterior diameter. (*D*) Lateral view one month prior to biopsy. Note mild restoration of vertebral height and absence of bone change in the subjacent eleventh dorsal body.

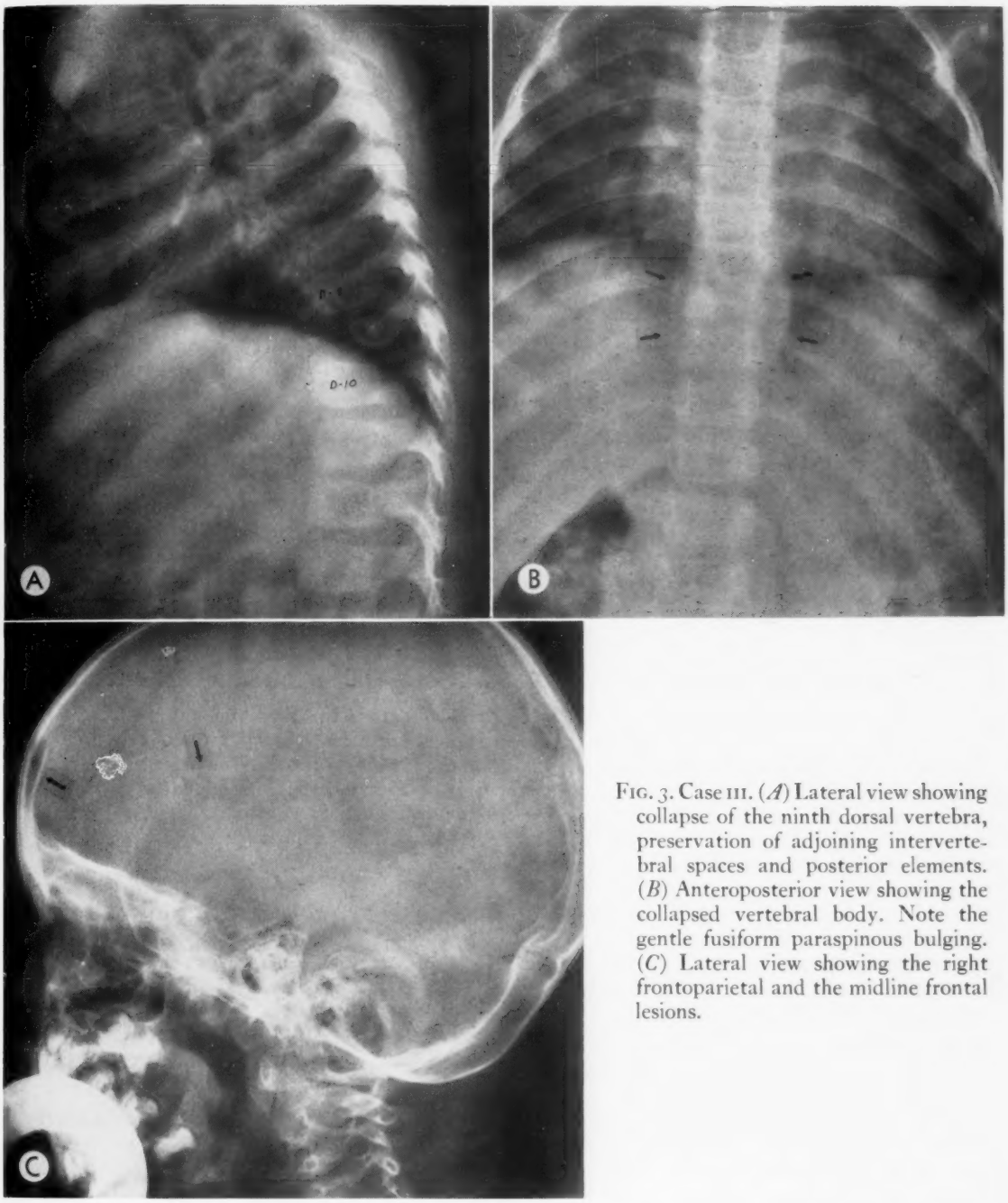


Fig. 3. Case III. (*A*) Lateral view showing collapse of the ninth dorsal vertebra, preservation of adjoining intervertebral spaces and posterior elements. (*B*) Anteroposterior view showing the collapsed vertebral body. Note the gentle fusiform paraspinous bulging. (*C*) Lateral view showing the right frontoparietal and the midline frontal lesions.

bar vertebra (Fig. 4 *A*), lytic areas in the proximal end of the left ulna (Fig. 4*C*), right temporoparietal region, right fifth rib, and left ilium.

*Laboratory Findings.* No significant abnormal findings.

*Operation.* A biopsy of the left ulnar lesion was performed.

*Pathologic Report.* Eosinophilic granuloma of bone (Dr. A. Nettleship).

*Course.* Deep roentgen therapy (half value layer 1.35 mm. Cu) was given to all areas. Each lesion received approximately 1,050 r to the skin with depth doses ranging from 630 r to 840 r. The child was followed in the clinic and soon became asymptomatic. All lesions rapidly healed after roentgen treatment (Fig. 4 *B*). A new lesion developed in the left eighth rib several months later and promptly healed after similar treatment.

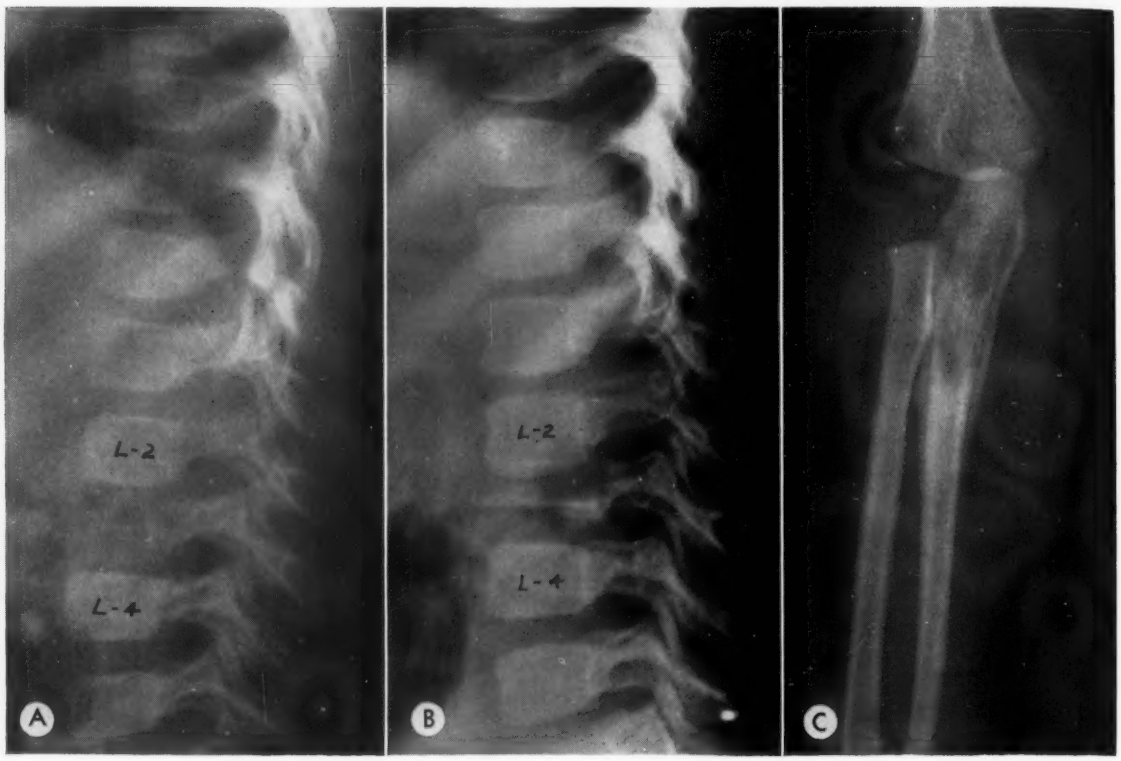


FIG. 4. Case IV. (*A*) Lateral view showing destruction and collapse of the third lumbar vertebral body and preservation of the adjoining intervertebral spaces and posterior elements. The paraspinous swelling cannot be seen in the anteroposterior view due either to its absence or obliteration by the psoas muscles or overlying intestinal gas shadows. (*B*) Lateral roentgenogram seven months after roentgen therapy. Note the mild degree of regeneration of the body, the preservation of the adjoining intervertebral spaces, and the increase in the anteroposterior diameter of the involved body. (*C*) An anteroposterior view showing the lytic area in the proximal shaft of the left ulna.

## DISCUSSION

We are convinced, as are Compere and his associates,[4] that the xanthomatous diseases can and will account for the vertebral deformity which has been termed "vertebra plana." Two cases in which we have biopsied the vertebral lesion itself have proved to be eosinophilic granuloma. We have found no cases with biopsy proof of osteochondritis or aseptic necrosis, although our search has not been exhaustive. On the other hand, cases have been reported with histologic proof of other conditions as was the case of malignant undifferentiated sarcoma cited by Dr. J. William Hillman in the floor discussion of the paper by Compere *et al.*[4] Possibly there is some common denominator, such as some roentgen or clinical feature, that might aid in making a preoperative clinical diagnosis. We would discourage the use of the term "vertebra plana" in the description of just any flattened vertebral body. If one would keep in mind the more important recurring roentgen and clinical findings, one could reasonably categorize those that would likely prove to be due to an underlying xanthomatosis. In this respect, the publication of others' experiences with proved cases might help to establish the criteria for preoperative diagnosis of this benign process which responds so readily to conservative measures.

## CONCLUSIONS

1. Xanthomatous diseases can and do produce vertebra plana.

2. Usually the clinical course is protean but can be very serious.

3. A mild bulging of the paraspinous shadows has been a consistent roentgen finding in our cases.

4. Roentgen therapy in moderate doses has proved to be of great value in these cases, even in one with severe neurologic disturbances.

Joseph D. Calhoun, M.D.
607 Donaghey Building
Little Rock, Arkansas

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# THE SITES OF MATURATIONAL STRENGTH AND WEAKNESS IN THE DEVELOPING HAND SKELETON*

*By* SAMUEL DREIZEN, M.D.,[1] RICHARD M. SNODGRASSE, PH.D.,[2] HAMILTON WEBB-PEPLOE,[3] JO G. DREIZEN, B.A.,[4] *and* TOM D. SPIES, M.D.[5]
BIRMINGHAM, ALABAMA

ASSESSMENT of skeletal maturity by the Todd[15] inspectional method involves the timing of the successive irreversible stages which specifically characterize the postnatal development of each articular surface and epiphyseal cartilage plate. In standards constructed for use with the Todd technique,[5,15] bone age, determined from the maturational progress of each ossification center in the hand and wrist, is conventionally equated with chronologic age in a 1:1 ratio. Theoretically, each ossification center should mature at the same rate; actually, complete symmetry in bone development is extremely rare. Greulich and Pyle[5] state that the bones of the hand and wrist are an aggregate of partially independent elements despite a tendency to act more or less as a unit in development.

Quantitatively, the degree of skeletal asymmetry throughout the growth period can be expressed by the red graph method.[10] In this procedure, the stage of development of each ossification center is identified in serial roentgenograms. As shown in Figure 1, the values ascribed to the most advanced and least advanced bones are plotted to delimit a zone which includes the maturities of all the other centers in the skeletal area. The sites of maturational strength and weakness at any period during the growth cycle of a child can thus be demarcated.

Asymmetry in bone development may appear very early in the course of skeletal maturation. Studies of the postnatal ossification centers in the hand and wrist show that the carpals are much more variable in time of appearance than the metacarpal, phalangeal, and distal radial and ulnar epiphyses.[2,7,11] In a previous report from this Clinic[2] in which the onset of ossification was investigated quantitatively, the greatest delay occurred in the greater multangular in both sexes. The present study was devised to determine the pattern of maturational dispersion during the subsequent phases of skeletal development and to establish the anatomic locations of the maturational extremes in the skeletal development of the hand in children as ethnically homogeneous as can be found in this country.

## MATERIALS AND METHODS

Serial roentgenograms of the left hand and wrist of 1,025 children in the growth study series of the Nutrition Clinic, Hillman Hospital, Birmingham, Alabama, were examined. The maturity of each of 31 ossification centers was assessed by the Todd[15] inspectional method using the Greulich-Pyle[5] "Radiographic Atlas of Skeletal Development of the Hand and Wrist" as the standard of reference. To facilitate analysis of the data, the assessments were screened for those which contained a single most advanced and a single least advanced ossification center. Those with two or more centers at either or both ends of the range of maturities were excluded. The final se-

* Northwestern University studies in nutrition at the Jefferson-Hillman Hospital, Birmingham, Alabama. From the Department of Nutrition and Metabolism, Northwestern University.
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[1] Assistant Professor of Nutrition and Metabolism, Northwestern University Medical School.
[2] Professor of Anatomy, Temple University School of Dentistry.
[3] Research Technician, Nutrition Clinic, Hillman Hospital.
[4] Research Associate, Department of Nutrition and Metabolism, Northwestern University Medical School.
[5] Professor of Nutrition and Metabolism, Northwestern University Medical School.

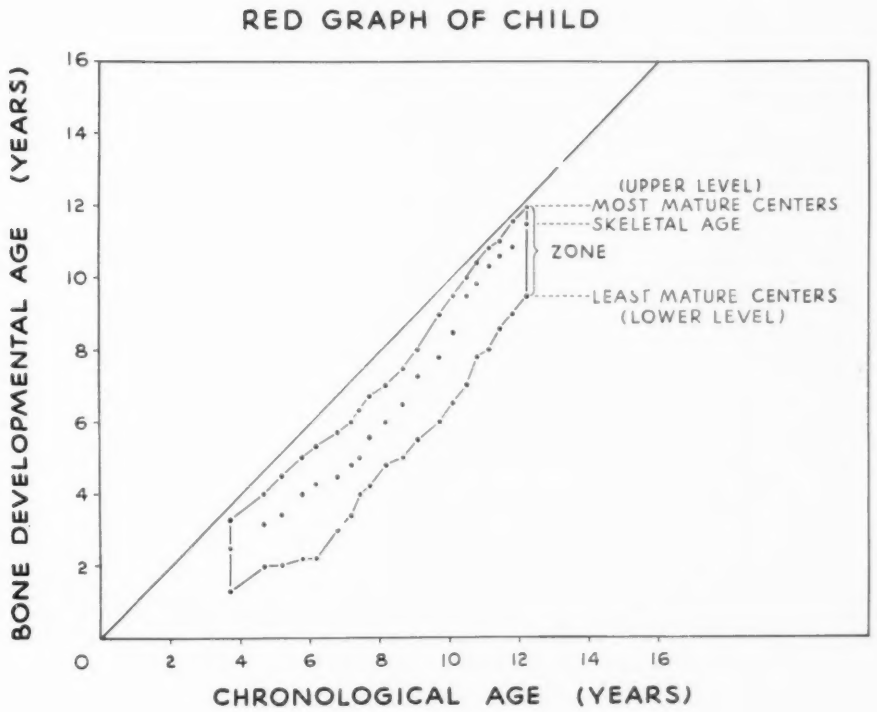


Fig. 1.

lection was restricted arbitrarily to one assessment per child. The arithmetic mean of the 31 maturities in the assessment comprised the skeletal age.

The criterion of a single most advanced and a single least advanced ossification center in a single roentgenogram was met by 740 children. Of these, 371 were boys and 369 girls. Each was white, of British extraction and a native of north-central Alabama. They ranged in age from seven months to eighteen years.

The frequencies with which each of the 31 primary and secondary postnatal ossification centers in the hand and wrist were rated as most mature or least mature in the 740 children were determined. Those for the carpals and adductor and flexor sesamoids of the thumb were adjusted in accordance with the age of appearance of these centers contained in the Greulich-Pyle[5] atlas. Assessment of the maturity status of the metacarpals, phalanges, and distal radius and ulna prior to the emergence of their respective epiphyses were based primarily on the contours of the articular surfaces and secondarily on the over-all size of the shafts. Frequencies were tabulated for each of 31 ossification centers in the hand and wrist and for the anatomic segments in which they were located. The influence of age, sex, and skeletal growth rate on the frequency distributions of the most advanced and least advanced centers was then studied.

## OBSERVATIONS

The anatomic dispersion of the most mature and least mature ossification centers in the hand skeletons of the 740 children is shown in Figures 2 and 3. For the group, the centers most frequently rated most mature were the triquetrum (112 cases, 15.1 per cent), distal radius (72 cases, 9.7 per cent), and distal ulna (68 cases, 9.2 per cent). The frequency scores for the remaining carpals exclusive of the pisiform greatly exceeded those of all metacarpals and thumb sesamoids and all but one of the phalanges. The frequency for distal phalanx I was of the same order of magnitude as that of most of the carpals. The pattern characteristic of

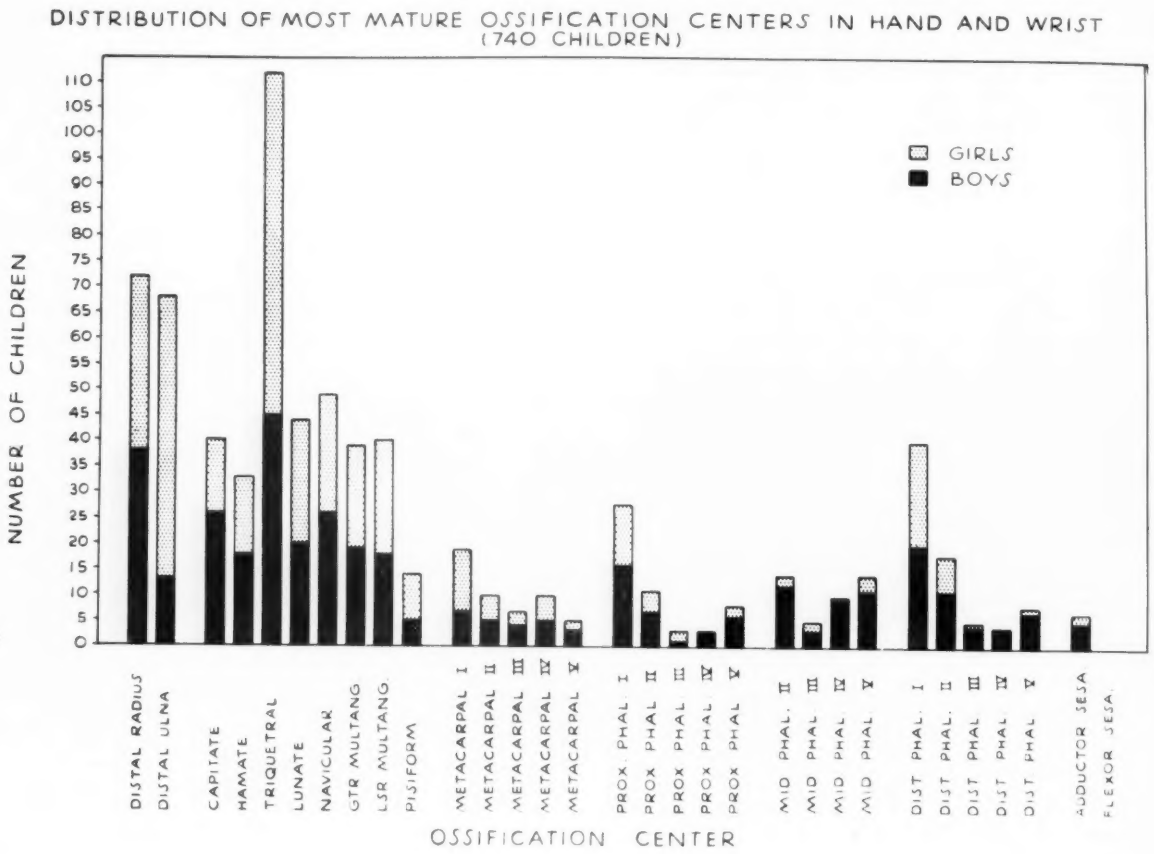


FIG. 2.

TABLE I

FREQUENCY SCORES FOR MOST MATURE AND LEAST MATURE PRIMARY OSSIFICATION CENTERS CORRECTED FOR AGE OF APPEARANCE IN ROENTGENOGRAM

| Center | Most Mature | | | | Least Mature | | | |
|---|---|---|---|---|---|---|---|---|
| | Boys | | Girls | | Boys | | Girls | |
| | Unc.* (per cent) | Cor.† (per cent) | Unc. (per cent) | Cor. (per cent) | Unc. (per cent) | Cor. (per cent) | Unc. (per cent) | Cor. (per cent) |
| Capitate | 7.0 | 7.0 | 3.8 | 3.8 | 4.6 | 4.6 | 5.4 | 5.4 |
| Hamate | 4.9 | 4.9 | 4.1 | 4.1 | 9.4 | 9.4 | 6.0 | 6.0 |
| Triquetrum | 12.1 | 12.8 | 18.1 | 18.9 | 5.9 | 6.2 | 5.4 | 5.6 |
| Lunate | 5.4 | 6.1 | 6.5 | 7.2 | 12.9 | 14.6 | 12.7 | 14.2 |
| Navicular | 7.0 | 9.8 | 6.2 | 7.8 | 3.0 | 4.2 | 8.7 | 10.6 |
| Greater Multangular | 5.1 | 6.3 | 5.4 | 6.6 | 6.0 | 7.9 | 6.8 | 8.2 |
| Lesser Multangular | 4.9 | 6.7 | 6.0 | 7.2 | 1.9 | 2.6 | 4.3 | 5.3 |
| Pisiform | 1.4 | 3.7 | 2.4 | 5.9 | 2.7 | 7.5 | 1.6 | 3.3 |
| Adductor Sesamoid | 1.4 | 8.9 | 0.5 | 2.8 | 0.3 | 1.8 | 0.0 | 0.0 |
| Flexor Sesamoid | 0.0 | 0.0 | 0.0 | 0.0 | 0.3 | 2.0 | 0.3 | 1.5 |

* Unc. = uncorrected.
† Cor. = corrected.

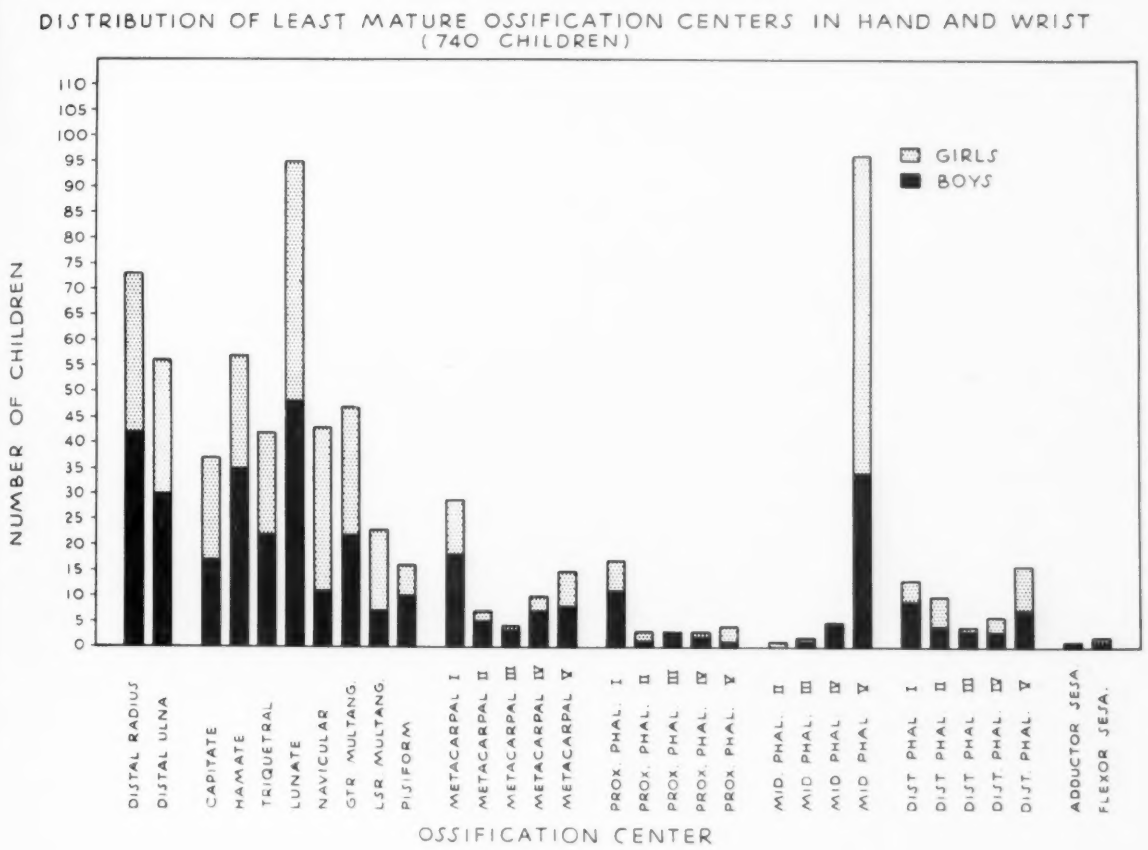


Fig. 3.

the group was sustained, in general, by the scores for the boys and girls separately. The most commonly advanced centers in the boys were the triquetrum (45 cases, 12.1 per cent) and distal radius (38 cases, 10.2 per cent); in the girls, the triquetrum (67 cases, 18.1 per cent) and distal ulna (55 cases, 14.9 per cent). As shown in Table I, adjustment of the data to accord with the time of appearance of the carpals and thumb sesamoids did not materially alter the relative distribution of the frequency scores except for the adductor sesamoid.

The most common least advanced centers for the entire group were the mesophalanx of the fifth digit (96 cases, 13.0 per cent), lunate (95 cases, 12.8 per cent), and distal radius (73 cases, 9.9 per cent) followed in descending order by the hamate, distal ulna, remaining carpals, metacarpals, phalanges and thumb sesamoids (Fig. 2 and 3). In the boys, the most prevalent least advanced centers were the lunate (48 cases, 12.9 per cent), distal radius (42 cases, 11.3 per cent), hamate (35 cases, 9.4 per cent), and middle phalanx V (34 cases, 9.3 per cent); in the girls, the middle phalanx V (62 cases, 16.8 per cent) and lunate (47 cases, 12.7 per cent). The basic distribution for each sex and for the group was essentially unchanged when the frequency scores for the carpals and thumb sesamoids were adjusted for the time of appearance of these bones (Table I).

Table II contains the groupings of the most mature and least mature centers in terms of their location in the various horizontal anatomic segments of the hand and wrist. The greatest frequencies for both the most advanced and least advanced bones occurred in the proximal and distal rows of carpals and in the distal radius and ulna combined. The metacarpals, proximal pha-

TABLE II

DISTRIBUTION OF MOST MATURE AND LEAST MATURE OSSIFICATION CENTERS BY ANATOMIC SEGMENTS

| Segment | Most Mature | | | | | | Least Mature | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | Boys | | Girls | | Total | | Boys | | Girls | | Total | |
| | No. | Per Cent | No. | Per Cent | No. | Per Cent | No. | Per Cent | No. | Per Cent | No. | Per Cent |
| Distal Radius and Ulna | 51 | 13.7 | 89 | 24.1 | 140 | 18.9 | 72 | 19.4 | 57 | 15.4 | 129 | 17.5 |
| Proximal Carpals | 96 | 25.9 | 123 | 33.2 | 219 | 29.5 | 91 | 24.5 | 105 | 28.4 | 196 | 26.6 |
| Distal Carpals | 81 | 21.9 | 71 | 19.3 | 152 | 20.5 | 81 | 21.9 | 83 | 22.5 | 164 | 22.2 |
| Metacarpals | 24 | 6.6 | 27 | 7.1 | 51 | 6.9 | 41 | 11.2 | 24 | 6.5 | 65 | 8.7 |
| Proximal Phalanges | 33 | 8.9 | 20 | 5.3 | 53 | 7.2 | 18 | 4.9 | 12 | 3.2 | 30 | 4.0 |
| Middle Phalanges | 35 | 9.3 | 8 | 2.1 | 43 | 5.8 | 40 | 11.0 | 64 | 17.4 | 104 | 14.0 |
| Distal Phalanges | 46 | 12.5 | 29 | 7.9 | 75 | 10.0 | 26 | 7.0 | 23 | 6.2 | 49 | 6.7 |
| Thumb Sesamoids | 5 | 1.4 | 2 | 0.5 | 7 | 0.9 | 2 | 0.6 | 1 | 0.3 | 3 | 0.4 |

langes and thumb sesamoids were found least often at either end of the range of maturities. When grouped on the basis of the digits involved, the first digit contained the highest number of most advanced centers and the fifth digit the greatest number of least mature centers.

The distribution of the anatomic sites of the most advanced and least advanced bone centers in relation to the rate of skeletal maturation is presented in Figures 4 and 5. Skeletal age equalled or exceeded chronologic age in 108 (14.6 per cent) children (58 boys and 50 girls). In 632 (85.4 per cent) children (313 boys and 319 girls), chronologic age surpassed skeletal age. The triquetrum was most frequently the farthest advanced center in each sex

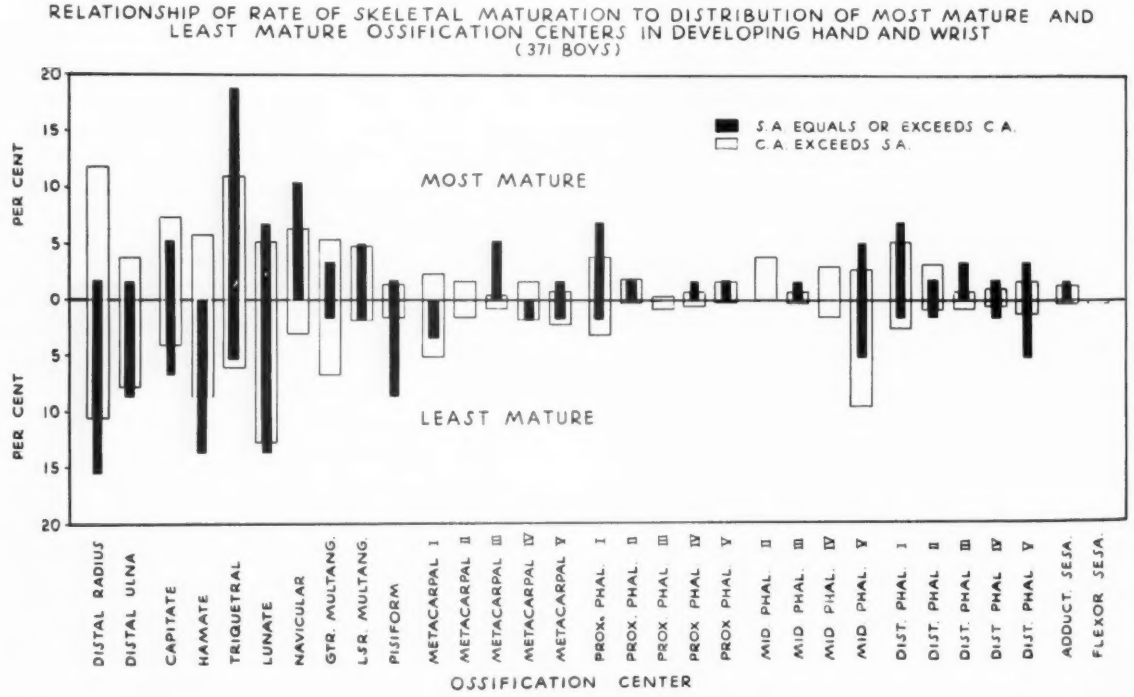


FIG. 4.

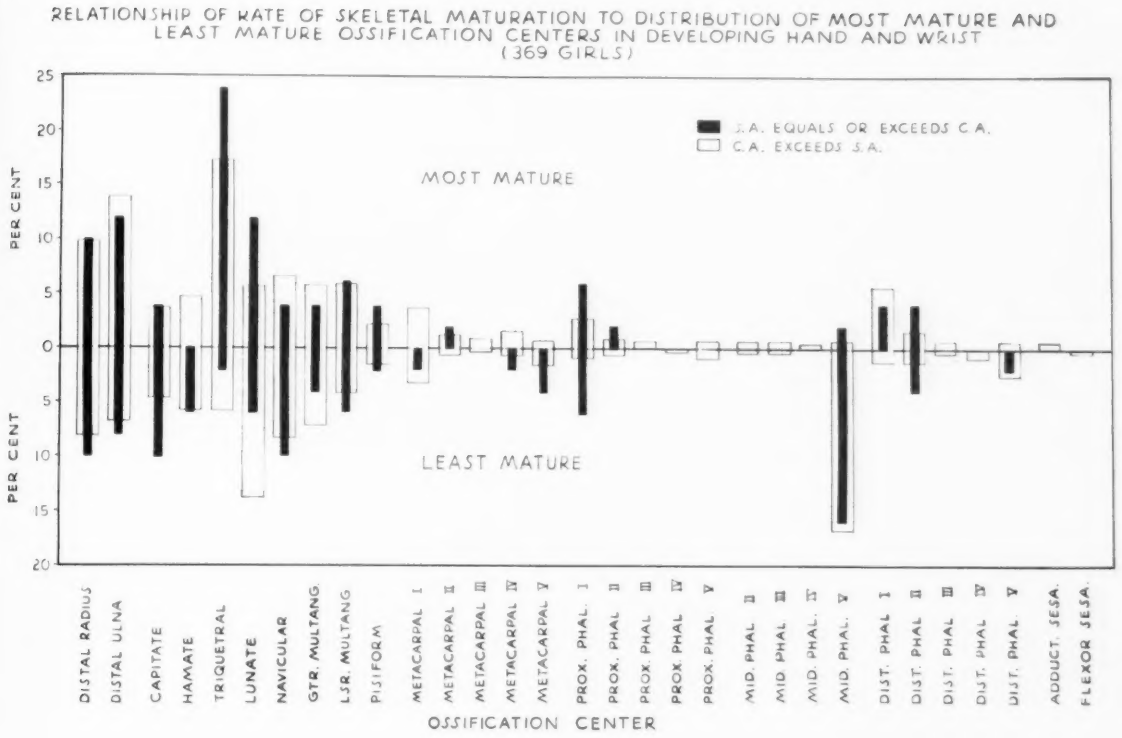


FIG. 5.

regardless of the speed of skeletal maturation. In contrast, the most common least advanced centers in the boys were the distal radius in those whose skeletal age equalled or exceeded chronologic age and the lunate in those whose chronologic age was greater than skeletal age. In the girls, the mesophalanx of the fifth digit was the most frequent least advanced bone in both the fast and slow maturing groups.

The Greulich-Pyle standard value for chronologic age was equalled by the most advanced center in 16 (5.1 per cent) and exceeded in 142 (45.3 per cent) of the 313 boys with a retarded over-all skeletal age. The corresponding distribution for the 319 girls with a lagging skeletal age were 7 (2.2 per cent) and 117 (36.4 per cent) respectively. As shown in Table III, the center which most frequently equalled or surpassed the chronologic age in the cases of retarded skeletal age was the triquetrum in the boys and the distal ulna in the girls. Conversely, the least mature center exceeded the standard for the corresponding chronologic age in only 20 (18.5 per cent) of the 108 children with a skeletal age which was either on par or advanced. Of the 20 cases, 4 were located in the distal radius and ulna, 9 in carpals, 1 in a metacarpal, 2 in middle phalanges and 4 in distal phalanges.

The frequency ratings for the three most common most advanced centers (triquetrum, distal radius and distal ulna) and for the three most common least advanced centers (middle phalanx V, lunate and distal radius) throughout the chronologic age range of the 740 children are shown in Table IV. The peaks of occurrence of the most advanced centers were for triquetrum between seven months and thirteen years, for distal radius between seven months and nine years and between thirteen years and eighteen years, and for distal ulna between five years and fifteen years. Those of the least advanced centers were for middle phalanx V between three years and twelve years, for lunate between five years and sixteen years, and for distal radius between

TABLE III

FREQUENCIES WITH WHICH MOST MATURE CENTER EQUALLED OR EXCEEDED STANDARD FOR CHRONOLOGIC AGE

| Bone | S.A.>C.A.* | | | S.A.=C.A. | | | S.A.<C.A. | | |
|---|---|---|---|---|---|---|---|---|---|
| | Boys | Girls | Total | Boys | Girls | Total | Boys | Girls | Total |
| Distal End of Radius | 0 | 4 | 4 | 1 | 1 | 2 | 16 | 12 | 28 |
| Distal End of Ulna | 1 | 6 | 7 | 0 | 0 | 0 | 9 | 21 | 30 |
| Capitate | 2 | 2 | 4 | 1 | 0 | 1 | 7 | 3 | 10 |
| Hamate | 0 | 0 | 0 | 0 | 0 | 0 | 10 | 4 | 14 |
| Triquetrum | 9 | 10 | 19 | 2 | 2 | 4 | 23 | 17 | 40 |
| Lunate | 4 | 4 | 8 | 0 | 2 | 2 | 3 | 10 | 13 |
| Navicular | 5 | 1 | 6 | 1 | 1 | 2 | 15 | 10 | 25 |
| Greater Multangular | 2 | 2 | 4 | 0 | 0 | 0 | 8 | 9 | 17 |
| Lesser Multangular | 3 | 2 | 5 | 0 | 1 | 1 | 12 | 5 | 17 |
| Pisiform | 1 | 2 | 3 | 0 | 0 | 0 | 1 | 6 | 7 |
| Metacarpal I | 0 | 0 | 0 | 0 | 0 | 0 | 3 | 6 | 9 |
| Metacarpal II | 0 | 1 | 1 | 0 | 0 | 0 | 2 | 1 | 3 |
| Metacarpal III | 3 | 0 | 3 | 0 | 0 | 0 | 1 | 1 | 2 |
| Metacarpal IV | 0 | 0 | 0 | 0 | 0 | 0 | 2 | 1 | 3 |
| Metacarpal V | 1 | 0 | 1 | 0 | 0 | 0 | 2 | 2 | 4 |
| Proximal Phalanx I | 4 | 2 | 6 | 0 | 0 | 0 | 4 | 2 | 6 |
| Proximal Phalanx II | 0 | 1 | 1 | 1 | 0 | 1 | 6 | 1 | 7 |
| Proximal Phalanx III | 0 | 0 | 0 | 0 | 0 | 0 | 1 | 0 | 1 |
| Proximal Phalanx IV | 1 | 0 | 1 | 0 | 0 | 0 | 2 | 0 | 2 |
| Proximal Phalanx V | 1 | 0 | 1 | 0 | 0 | 0 | 3 | 1 | 4 |
| Middle Phalanx II | 0 | 0 | 0 | 0 | 0 | 0 | 7 | 1 | 8 |
| Middle Phalanx III | 1 | 0 | 1 | 0 | 0 | 0 | 0 | 1 | 1 |
| Middle Phalanx IV | 0 | 0 | 0 | 0 | 0 | 0 | 5 | 0 | 5 |
| Middle Phalanx V | 3 | 1 | 4 | 0 | 0 | 0 | 3 | 1 | 4 |
| Distal Phalanx I | 4 | 1 | 5 | 0 | 2 | 2 | 9 | 5 | 14 |
| Distal Phalanx II | 1 | 1 | 2 | 0 | 1 | 1 | 2 | 2 | 4 |
| Distal Phalanx III | 1 | 0 | 1 | 1 | 0 | 1 | 1 | 1 | 2 |
| Distal Phalanx IV | 1 | 0 | 1 | 0 | 0 | 0 | 1 | 0 | 1 |
| Distal Phalanx V | 1 | 0 | 1 | 1 | 0 | 1 | 0 | 0 | 0 |
| Adductor Sesamoid (Thumb) | 1 | 0 | 1 | 0 | 0 | 0 | 0 | 1 | 1 |
| Flexor Sesamoid (Thumb) | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 |
| Total | 50 | 40 | 90 | 8 | 10 | 18 | 158 | 124 | 282 |

* S.A.=Skeletal Age.
C.A.=Chronologic Age.

seven months and four years and between ten years and fourteen years.

The mean amount of deviation, in months, from the Greulich-Pyle standards for each of the most advanced and least advanced centers is depicted in Figures 6 and 7. Separate means were computed for assessments which exceeded the standard and for those which fell below the standard. The means for the most advanced centers with maturity ratings above the standard ranged from 1.5 months (metacarpal IV) to 15.3 months (middle phalanx V) advanced in the boys and from 1.0 month (metacarpal III, middle phalanx III and distal phalanx III) to 13.8 months

TABLE IV

FREQUENCIES OF OCCURRENCE OF THE THREE MOST COMMON UPPERMOST CENTERS AND THREE MOST COMMON LOWERMOST CENTERS AT VARIOUS CHRONOLOGIC AGES

| Chronologic Age (mo.) | No. of Children | Most Mature Centers | | | Least Mature Centers | | |
|---|---|---|---|---|---|---|---|
| | | Triquetrum (per cent) | Distal Radius (per cent) | Distal Ulna (per cent) | Middle Phalanx V (per cent) | Lunate (per cent) | Distal Radius (per cent) |
| 7–11 | 6 | 16.7 | 16.7 | 16.7 | 0.0 | 0.0 | 16.7 |
| 12–23 | 29 | 31.0 | 13.8 | 3.4 | 6.9 | 0.0 | 10.3 |
| 24–35 | 48 | 20.8 | 14.6 | 0.0 | 10.4 | 0.0 | 14.6 |
| 36–47 | 61 | 18.0 | 8.2 | 3.3 | 14.7 | 1.6 | 13.1 |
| 48–59 | 48 | 27.1 | 8.3 | 0.0 | 14.6 | 12.5 | 4.2 |
| 60–71 | 68 | 18.8 | 11.8 | 10.3 | 17.6 | 20.6 | 6.0 |
| 72–83 | 62 | 16.1 | 6.5 | 12.9 | 17.7 | 17.7 | 4.6 |
| 84–95 | 70 | 10.0 | 11.4 | 18.6 | 11.4 | 25.7 | 10.0 |
| 96–107 | 71 | 15.5 | 12.7 | 12.7 | 19.7 | 16.9 | 5.6 |
| 108–119 | 61 | 14.8 | 6.6 | 16.1 | 11.5 | 14.7 | 4.9 |
| 120–131 | 59 | 10.1 | 3.5 | 10.4 | 15.5 | 10.4 | 26.2 |
| 132–143 | 36 | 13.9 | 0.0 | 2.7 | 19.4 | 5.6 | 16.7 |
| 144–155 | 35 | 11.4 | 5.7 | 9.3 | 2.9 | 14.3 | 11.4 |
| 156–167 | 22 | 4.6 | 9.1 | 15.9 | 0.0 | 18.2 | 13.7 |
| 168–179 | 17 | 5.5 | 11.8 | 11.8 | 6.0 | 11.8 | 0.0 |
| 180–191 | 14 | 0.0 | 13.3 | 0.0 | 0.0 | 28.4 | 7.0 |
| 192–203 | 23 | 4.3 | 21.8 | 4.3 | 8.7 | 0.0 | 0.0 |
| 204–216 | 10 | 0.0 | 30.0 | 0.0 | 0.0 | 0.0 | 0.0 |

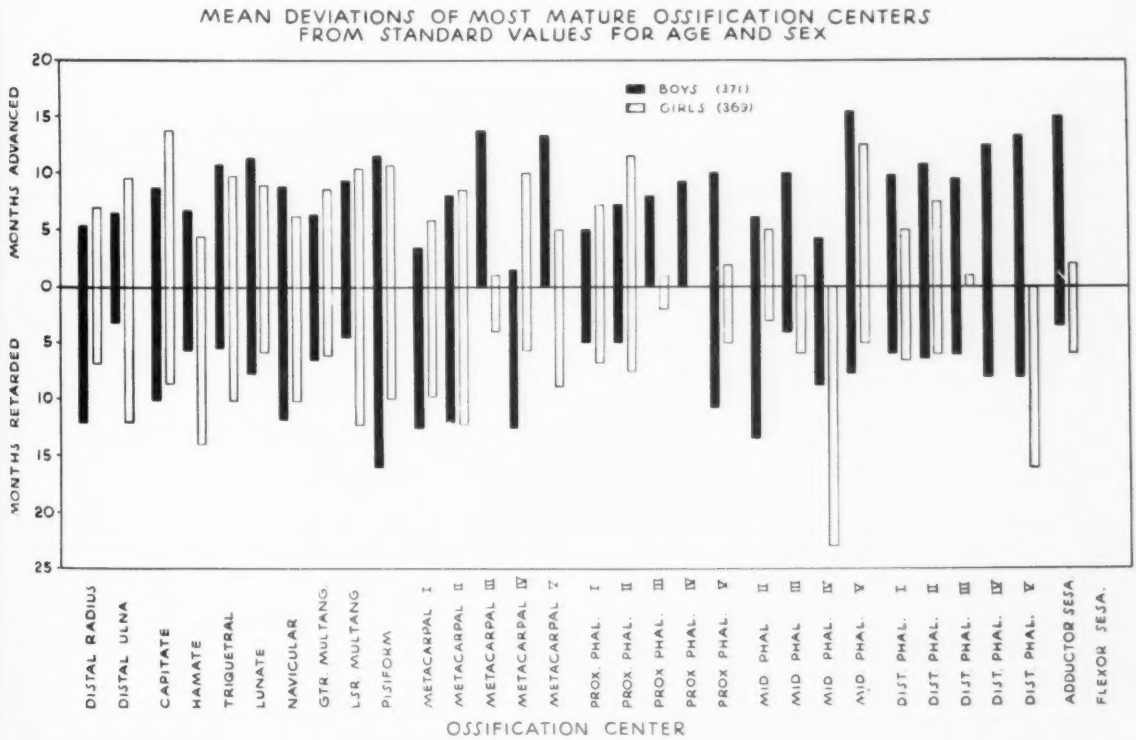


FIG. 6.

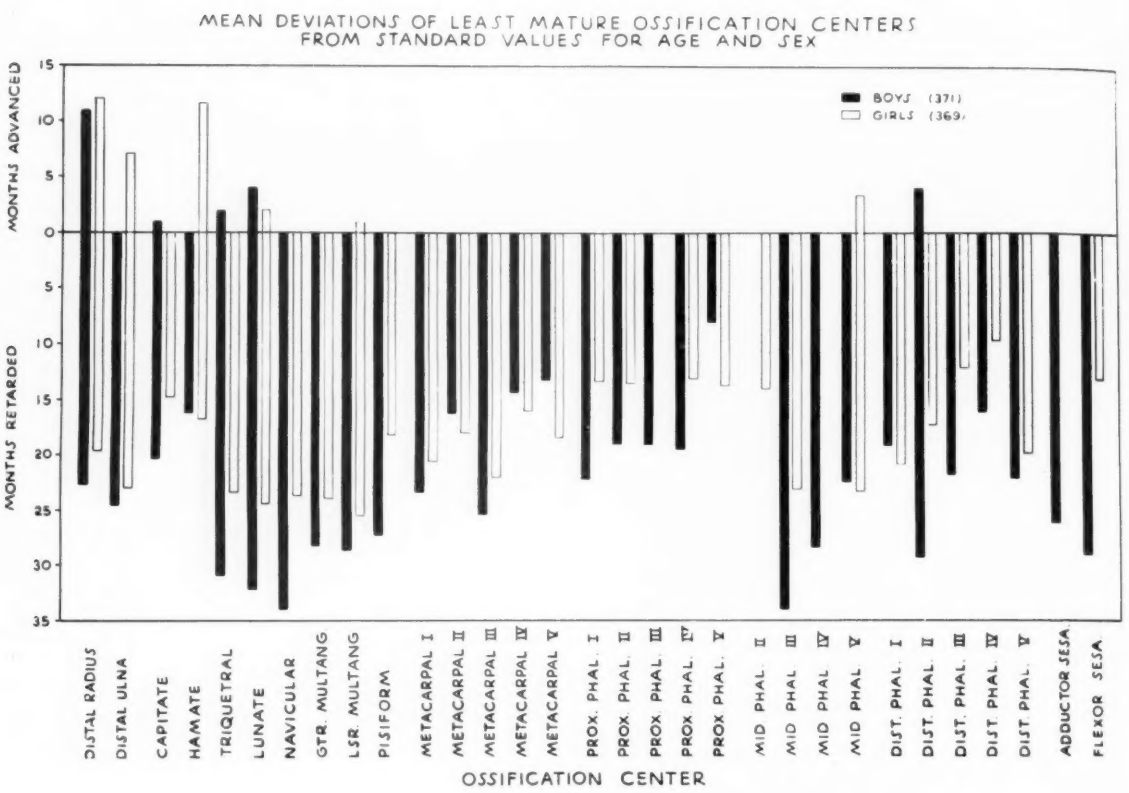


FIG. 7.

(capitate) advanced in the girls. Where the maturities of the most advanced centers were below standard, the means extended from 3.3 months (distal ulna) to 16.0 months (pisiform) retarded in the boys and from 2.0 months (proximal phalanx III) to 23.0 months (middle phalanx IV) retarded in the girls. In the cases in which the maturities of the least advanced centers exceeded the standard, the range extended from 1.0 month (capitate) to 11.0 months (distal radius) advanced in the boys and from 1.0 month (lesser multangular) to 12.0 months (distal radius) advanced in the girls. The corresponding ranges for the least mature centers below standard were 8.0 months (proximal phalanx V) to 34.0 months (navicular and middle phalanx III) retarded in the boys and from 9.7 months (distal phalanx IV) to 25.4 months (greater multangular) retarded in the girls.

## DISCUSSION

The demonstration that certain ossification centers in the developing hand skeleton occur with unusual frequency at either end of the range of maturities is indicative of the individuality of the component parts of this skeletal unit. Variations in maturity among the bones of the growing hand were attributed by Todd *et al.*[15] to the differential vulnerability of these centers. In recognition of this differential, Todd and his coworkers[15] emphasized that the greatest reliance in estimating bodily maturity be placed on the metacarpal and phalangeal epiphyses. The carpals, particularly the triquetrum, and the distal radial and ulnar epiphyses were regarded as being exceptionally susceptible to disturbances in the ossification process and therefore much less dependable as determinants of over-all maturity than the metacarpals and phal-

anges. Milman and Bakwin[9] found that because of their relative stability, the epiphyses of the first and second metacarpals were the bones in the hand most suitable as maturity indicators for clinical purposes. The present findings that the triquetrum, distal radius, and distal ulna were the most common sites of greatest maturity and that the lunate and distal radius were two of the three most common locations of least maturity are in general agreement with the concept that the carpals and distal radius and ulna are more variable in their maturational progress than the metacarpals and phalanges.

The triquetrum was the most frequent location of greatest maturational strength in both sexes and in both the fast and slow maturing groups in this series up to thirteen years of age. In the well-born, well-circumstanced and well-cared-for Cleveland children of North European ancestry studied by Todd and associates,[15] the triquetrum was the center most sensitive to disturbances in the ossification process and most likely to be lacking in infancy. Once lacking, it was most delayed in its appearance. Sontag and Lipford[14] hold that the chief cause of "aberrant" centers, *i.e.*, those appearing relatively early or late, is genetic; whereas Francis[3,4] implies that aberrant centers are primarily a reflection of a greater responsiveness to environmental stresses. The precise genetic and environmental factors contributing to the differences in the status of the triquetrum in the Alabama and Cleveland children are not irrefutably discernible. The magnitude of the difference underscores, however, the individuality of each center in the same child and in different children of similar and dissimilar ethnic backgrounds.

As the most common least mature ossification center in the 740 children, the mesophalanx of the fifth finger was unique among the seemingly less fluctuant phalanges and metacarpals in its maturational irregularity. The selective specificity of middle phalanx V to disturbances in ossification has been noted repeatedly in health and disease. Caffey[1] states that the middle and terminal phalanges of the fifth digit are hypoplastic in approximately 1 per cent of normal children. Shortening and malformation of the diaphysis of middle phalanx V has come to be regarded as a stigma of mongolism[6,12,13] and has been found occasionally in children with progressive myositis ossificans and psoriasis.[8] This anomaly has been described in clinically healthy children and is apparently more common in those with retarded skeletal maturation.[16,17]

## SUMMARY AND CONCLUSIONS

The sites of maturational strength and weakness in the developing bones of the hand and wrist were determined in 740 children from skeletal age assessments containing a single most mature and single least mature center. The influences of sex, chronologic age, and speed of skeletal maturation were evaluated in appropriate subgroups. It was found that:

1. The triquetrum, distal radius, and distal ulna were the most mature centers in 252 (34.0 per cent) of the 740 children; whereas the middle phalanx V, lunate, and distal radius were the least mature centers in 264 (35.7 per cent). The triquetrum was the most frequent most advanced center in both boys (12.1 per cent) and girls (18.1 per cent). In boys the most frequent least advanced center was the lunate (12.9 per cent), in girls the middle phalanx V (16.8 per cent).

2. Save for the mesophalanx of the fifth digit, the maturities of the phalanges and metacarpals showed a greater tendency to cluster around skeletal age (the arithmetic mean) than did the carpals and distal radius and ulna.

3. The three most advanced and three least advanced bones showed considerable overlapping in frequency throughout the chronologic age range of seven months to eighteen years.

4. The distribution of most advanced

and least advanced centers was not related to the over-all speed of skeletal maturation.

It is thus apparent that variations in the maturities of the 31 bones of the hand skeleton included in the skeletal age assessments are a reflection of individual bone response to the same and different degrees of disturbances in the ossification process.

Tom D. Spies, M.D.
Nutrition Clinic
Hillman Hospital
Birmingham, Alabama

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VOL. 82, NO. 3

# ROENTGEN CHANGES IN SICKLE CELL ANEMIA*

*By* KIRK R. DEIBERT, M.D.
FLORENCE, ALABAMA

SICKLE cell anemia is almost exclusively limited to the Negro race. Its manifestations are diffuse so that no tissue or viscus is spared from its involvement in the human host. Thus, this has stimulated a marked interest in those clinicians and radiologists who practice in the south. This presentation deals with an analysis of the clinical and roentgenologic aspects of 16 case studies.

## REVIEW OF THE LITERATURE

A review was made of approximately 30 articles or their abstracts which have appeared in the literature in the past ten years, some of them being listed in the bibliography of this paper.[1–6] In only one article was mention made of the bizarre flattening and broadening of the thoracic and lumbar vertebrae which were noted in adult Negroes. Numerous authors observed biconcave disk impressions upon the vertebral body plates. Abdominal distention was mentioned as a frequent finding in the typical acute sickling crisis, but not specifically an adynamic ileus, the pattern of which is illustrated in Figure 2. Many descriptions are also given of the typical osseous changes found on skull roentgenograms; however, in this survey the change most constantly observed was a coarse granular, somewhat porous appearance of the calvarium. This was noticeable especially in the midparietal area (Fig. 3*a*). The changes in the other bones, as obtained from routine roentgenographic survey study, varied in direct proportion to the number of authors. In this group, chiefly a generalized coarsened porous trabeculation pattern was noted. Quite frequently, faint linear oblique, vertical or parallel dense bands were identified at the endosteal medullary junction or at the metaphyseal diaphyseal junction.

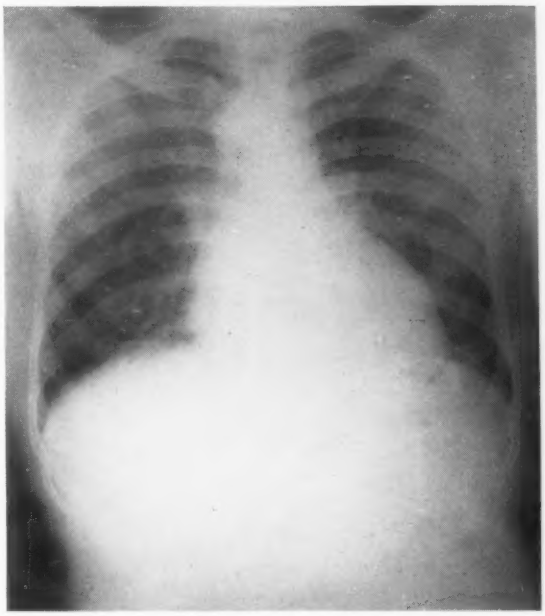

FIG. 1. Chest roentgenogram shows cardiomegaly, elevated right hemidiaphragm, bilateral basilar pleuropulmonary bands of diffuse pneumonitis and atelectasis.

## ROENTGEN FINDINGS

The roentgen findings which were observed in this series are presented and the roentgenograms of one typical patient with sickle cell anemia, who had a complete survey study, are reproduced (Fig. 1–4).

A. *Chest.* Cardiac enlargement was the most common finding. This was generalized and, if it was associated with a prominent outflow tract, rheumatic heart disease was often erroneously assumed to be present. Not infrequently, various pleuropulmonary densities were identified, compatible with pneumonitis, infarct or linear atelectasis. Changes of bone density and of the texture of the trabeculations were noted in the thoracic osseous structures. A slightly elevated hemidiaphragm, which was suggestive of hepatomegaly, was also seen (Fig. 1).

* Presented at the Third Annual Meeting of the Southern Radiological Conference, Point Clear, Alabama, January 31, 1959.

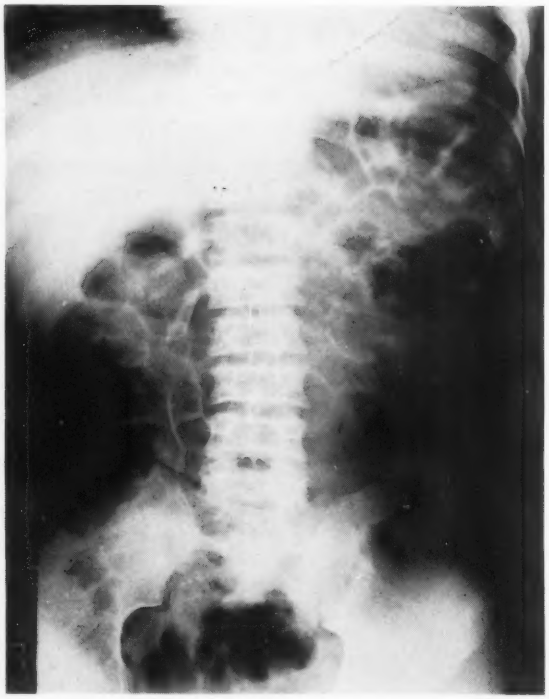

FIG. 2. Plain roentgenogram of the abdomen shows typical findings of sickle cell anemia: cardiomegaly, hepatomegaly, small bowel ileus reaction pattern coincident to a sickling crisis, coarse bone architecture of the right ilium, and diffuse vertebra plana with height-width ratio of 1:2.8.

B. *Abdomen.* A small bowel ileus occurred quite frequently (Fig. 2). Changes of pelvic bone density and architecture or aberrations of the vertebral body contour were found. Lienohepatomegaly and cholelithiasis were observed as well as abnormalities of the hip (Fig. 3*b*).

C. *Skull.* A fairly diffuse granular appearance was identified throughout the calvarium. This impression was chiefly gained from the mottled porosity of the diploic architecture (Fig. 3*a*). Recourse to a fourth power magnification hand lens will demonstrate this finding to better advantage.

D. *Peripheral Bone Structure.* The osseous changes which were visualized were quite variable and are considered in the order of importance: (1) a generalized change in bone architecture; namely, irregularity of the endosteum, diffuse porosis, faint linear or oblique dense bands, localized zones of radiolucency or in contrast sclerosis; (2) growth arrest lines; (3) periosteal elevation; (4) disturbed cortex medullary ratio; and (5) in the small bones, as the phalanges, a diffuse porosis, which is in sharp contrast to the disturbed pattern of the residual striae. This is not unlike that noted in hypothyroidism (Fig. 4).

E. *Joints.* Epiphysitis and aseptic necrosis were not infrequent findings. This was especially true of the hip as a weight bearing joint, resulting in coxa plana which progressed rapidly into a marked adult degenerative arthritis. The changes constituted an exact duplication of those seen in Legg-Perthes disease; however, in sickle cell anemia the cause is vascular throm-

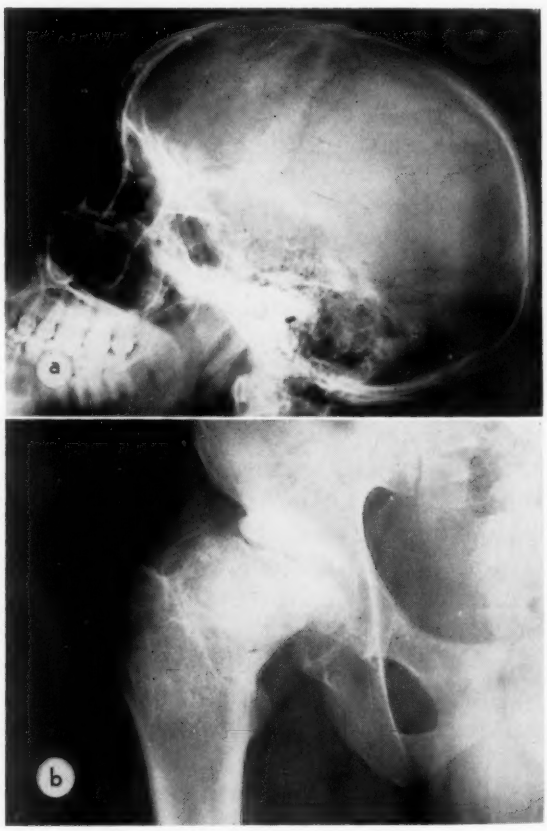


FIG. 3. (*a*) Lateral skull roentgenogram shows a diffuse granular porous diploic pattern. (*b*) Right hip roentgenogram shows capital epiphyseal aseptic necrosis and diffuse coarsening of femoral trabeculations. A secondary degenerative acetabular arthritis is noted.

bosis of the capital femoral epiphysis (Fig. 3*b*).

F. *Vertebra.* Reduction of the vertebral height and broadening of its transverse diameter with biconcave plate impressions were common findings. However, the recognition of a diffuse vertebra plana of the Calvé type should not be overlooked nor misinterpreted. It is to be noted that in an average adult a correlation exists between the vertical height and transverse breadth of a lumbar vertebra. The average height-width ratio is 1:1.7; in the case shown in the roentgenograms, it was 1:2.8 (Fig. 2).

## CLINICAL FINDINGS

The pathologic background is not discussed, since this has been covered by many previous authors; however, some of the typical clinical aspects of sickle cell anemia are enumerated. The list has been compiled from the clinical charts of the 16 patients. One case is deleted because the clinical laboratory studies revealed a thrombocytopenic aplastic anemia. Two patients had the typical roentgen changes attending sickle cell anemia but no twenty-four hour "wet-blood" preparation was made by the laboratory; therefore, they are also omitted. The remaining 13 patients had positive sickling of the red blood cells. In the order of importance and frequency of occurrence, the clinical findings were as follows: (1) anemia; (2) fever (102° F.); (3) leukocytosis (average 17,000); (4) bone and joint pain; (5) abdominal pain; (6) back pain; (7) cardiac enlargement; (8) skin lesions; (9) limp and scoliosis; (10) obstetric difficulty; and (11) visceromegaly.

## COMMENT

All of the patients had an anemia with an average red blood cell count of $2.2 \times 10^6$ and an average hemoglobin recording of 65 per cent. There was no relationship between the degree of anemia and the severity of the clinical symptoms.

Alternate periods of remission and recrudescence constitute the typical clinical course. Usually a patient is admitted to the hospital for weakness and malaise. Anemia is found and for relief hematinics and transfusions are prescribed. With progression and worsening of the patient's general state, a typical crisis may develop. At the peak of one of these violent cyclic attacks, the patient may experience pain anywhere in the body, mimicking almost any clinical syndrome. This leads to various roentgen examinations wherein a cooperative conference between clinician and radiologist is a requisite. The dictum, "to think of sickle cell anemia is to diagnose it," certainly holds true.

It has been noted that in a sickling crisis

Fig. 4. Roentgenograms of extremities in sickle cell anemia. (*a*) Right lower leg. Dense linear bands at metaphyseal and cortical endosteal junctions of tibia. Reduced fibular corticalis. (*b* and *c*) Right hand and ankle. Reduced cortical thickness causing relative broadening of the medulla with generalized porosity that accentuates the trabeculations.

attack erythrocytolysis becomes acute, being associated with leukocytosis, fever, pain at various sites and cardiac insufficiency. When the abdominal pain is severe and an adynamic ileus is visualized on a plain roentgenogram of the abdomen, the erroneous diagnosis of an acute emergent abdomen may be made (Fig. 2). Frequently, a crisis is precipitated by pneumonitis. Occasionally, it is difficult to distinguish pneumonitis from atelectasis or pulmonary infarction. Arterial thromboses are the cause of these varied pleuropulmonary densities. A similar cause has been found pathologically for the bizarre bone densities as well as for epiphyseal osteochondritis.

## DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes almost the entire gamut of the syndromes of clinical medicine. Among these are: (1) osteochondritis, Legg-Perthes and Calvé disease; (2) anemia, Cooley's type or hemolytic erythroblastic anemia; (3) hypothyroidism, cretinism or Cushing's syndrome; (4) Gaucher's or caisson disease; (5) leukemia, hemangioma or osteomyelitis; (6) xanthomatosis, neurofibromatosis, senile osteoporosis; (7) chondrodystrophia or lues; and (8) rheumatic disease.

## SUMMARY

The cardinal roentgen changes that should make one suspect sickle cell anemia are cardiomegaly, small bowel ileus, bizarre bone porosis and coarsened trabecular architecture, perhaps associated with the end result of aseptic necrosis.

When these findings are combined with the salient clinical aspects of anemia in a Negro, *i.e.*, fever, leukocytosis, bone joint and abdominal pains, the diagnosis of sickle cell anemia should be apparent.

Box 22, Rt. 1
Jackson Highway
Florence, Alabama

Acknowledgment is made to Dr. Loren Gary, Jr., 121 E. 5th St., Tuscumbia, Alabama, for permission to abstract clinical material and to reproduce roentgenograms (Fig. 1–4) of patient J. S., N.m. 20., and to Mr. Lewis D. Nichols, Department of Medical Illustration, Vanderbilt University Medical School, Nashville, Tennessee, for the photographic reproductions of the roentgenograms.

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VOL. 82, NO. 3

# THE ROENTGEN FINDINGS IN TRAUMATIC RUPTURE OF THE SPLEEN IN CHILDREN*

*By* SOLOMON S. SCHWARTZ, M.D., SCOTT J. BOLEY, M.D., *and* WILLIAM M. P. McKINNON, M.D.
BROOKLYN, NEW YORK

ARE the roentgen findings usually attributed to traumatic rupture of the spleen applicable to children? A unique opportunity to study this question recently presented itself during a review of 33 cases of this type of injury in children.[2]

Plain roentgenograms of the abdomen were available in 12 cases and were reviewed with respect to findings reported in the literature.[4–8,10,11] Table I lists these findings and their incidence in our series. Table II lists additional roentgen manifestations present in our cases and possibly related to ruptured spleen. In order to test the specificity of both groups of roentgen findings, it was decided to review plain roentgenograms of the abdomen of 200 infants and children. Most of these children were normal and the remainder suffered from conditions other than splenic rupture. Each finding will be discussed with reference to occurrence and significance in the study and control groups.

TABLE I

INCIDENCE OF ROENTGENOLOGIC FINDINGS PREVIOUSLY DESCRIBED IN RUPTURED SPLEEN

| | No. |
|---|---|
| A. Stomach | |
| 1. Exaggerated mucosal folds on greater curvature of stomach | 7 |
| 2. Gastric dilatation | 5 |
| 3. Increased gastrodiaphragmatic space | 0 |
| 4. Stomach displaced to right | 2 |
| B. Spleen | |
| 1. Obliteration of splenic shadow | 1 |
| 2. Increase in distance between kidney and spleen | 0 |
| 3. Enlargement of splenic shadow | 5 (2) |
| C. Pleural Reaction at Left Base | 0 |
| D. Rib Fracture | 0 |
| E. Bowel | |
| 1. Widening of space between bowel loops | 0 |
| 2. Distention of both large and small bowel | 1 |
| 3. Depressed splenic flexure | 1 |
| F. Obliteration of Various Shadows | |
| Kidney | 0 |
| Psoas | 0 |
| Abdominal haze | 0 |

## ROENTGEN FINDINGS PREVIOUSLY DESCRIBED

### A. STOMACH

1. *Exaggerated Mucosal Folds on Greater Curvature of Stomach.* This was present 7 times in our 12 cases and in 6 per cent of controls. It is said to be due to dissection of blood along the course of the vasa brevia. Mucosal folds may vary in shape and size normally. The shape changes with activity of the muscularis mucosae, perhaps directly by effect of contraction but more likely as a result of change in volume of tissue fluid with contracture. The size also varies with tissue fluid but may be a manifestation of increase in cellular content of the folds. It is thus difficult to attribute pathologic significance to something that may occur physiologically. Despite its occurrence in other conditions, this was the most common single roentgen finding in our series and therefore serves as a warning beacon.

2. *Gastric Dilatation.* This was present in 5 cases and in 3 per cent of controls. The stomach, like any other gas-containing, hollow viscus, may respond to any nearby irritant, such as blood, by reflexly dilating. Gastric distention normally occurs following ingestion of a large meal. Therefore, dilatation must be interpreted with respect to the time of last eating. If this history is not available, then distention must be

* From the Department of Radiology and Pediatric Surgical Division, Jewish Hospital of Brooklyn and the Department of Surgery, Queens Hospital Center.

TABLE II

INCIDENCE OF ADDITIONAL ROENTGENOLOGIC FINDINGS NOTED IN THIS SERIES

| | No. |
|---|---|
| A. Increase in Gastrocolic Space | 2 |
| B. Scoliosis to Right | 4 |
| C. Sentinel Distention of Transverse Colon or Distention of Colon to Splenic Flexure | 3 |
| D. Obliteration of Left Properitoneal Fat Line | 2 |

shown to persist beyond a two to three hour period, when a dilated but normally active stomach would be expected to empty itself.

3. *Increased Gastrodiaphragmatic Space.* This is supposed to be due to blood collecting between the fundus and the diaphragm or to pleural reaction secondary to diaphragmatic irritation. No obvious increase in this space was present in any of our cases. In reviewing the control series it became apparent that this finding cannot be evaluated on the supine roentgenogram of the abdomen. With the patient supine, the fundus is normally dependent and fluid migrates into it. Thus, any measurement from diaphragm to gas bubble will be increased by that portion of the fundus obscured by fluid. The measurement should be made, therefore, only in the erect position where air rises into the fundus, establishing the true upper gastric border. Further, the measurement made in the frontal projection, even in the erect position, is frequently invalid because the diaphragm, especially in children, rises to a considerably higher level anteriorly than posteriorly. The anterior portion of the diaphragm is thus projected above the gas bubble. The true situation is revealed only in an erect lateral view.

4. *Stomach Displaced to Right.* This occurred twice in our series and in 5 of the controls. In both cases with splenic rupture subcapsular hemorrhage was responsible and also produced gross roentgen enlargement of the splenic shadow. In the controls displacement was due to enlargement of the left kidney in 3 cases, 2 with Wilms' tumor and 1 with a large unilocular cyst. A fourth control had splenomegaly secondary to leukemia and the other had splenodiaphragmatic interposition of the colon. It is apparent that any space-taking lesion in the left upper quadrant is capable of producing this sign. In theory, traction on the lesser curvature of the stomach can do the same. Gastric displacement is relatively uncommon because significant consumption of left upper quadrant space following splenic trauma seldom occurs. This is discussed further under the section on changes of splenic shadow.

B. SPLEEN

1. *Obliteration of Splenic Shadow.* Since a splenic shadow was not visualized in 73 per cent of controls, it is difficult to talk of obliteration of this shadow. Obscuration of the splenic shadow is, therefore, interpreted to mean an ill-defined left upper quadrant mass and was present in 1 of our cases.

Of interest is the finding that the younger the child, the less frequently was a splenic shadow seen. This is attributed to a lack of perisplenic fat. In 3 of the 4 infant controls where the spleen was seen, the shadow was present on some roentgenograms and not on others, varying with phase of respiration. On deep inspiration the spleen is pushed against and contrasted with the air-containing portion of the stomach. Therefore, the introduction of gas into the stomach may be used to visualize a splenic shadow not seen on the plain roentgenogram of the abdomen.

2. *Increase in Distance between Kidney and Spleen.* To our knowledge no normal measurements for this distance have been established. Further, the equal difficulty in identifying the splenic and renal shadows in this age group would make any such measurement inaccurate.

3. *Enlargement of Splenic Shadow.* Under this heading, we include two groups of cases, those with gross enlargement of the splenic shadow and those with the tip of

the spleen presenting beneath the twelfth rib. Two of our cases and 5 controls fall into the former category; 3 of our cases and 18 controls fall into the latter.

Of the 7 cases with gross enlargement of the splenic shadow, the 2 with splenic injury had subcapsular bleeding and the 5 controls had true splenomegaly. While perisplenic clots did not produce this sign in our series, it has been shown to do so in adults. Therefore, the presence of such gross enlargement must be interpreted as either true splenomegaly or intra- or perisplenic bleeding. As spleens enlarge, the danger of rupture following minimal trauma increases. In view of this, when a patient with clinical evidence of splenic injury presents with gross enlargement of the splenic shadow, exact roentgen interpretation is not possible. If the enlargement is attributed to traumatic rupture with perisplenic or subcapsular hemorrhage, the correct clinical diagnosis may have been made for the wrong reason as at operation rupture of an enlarged spleen may be found. In such a case, the change in size of splenic shadow is a reflection of splenomegaly and not of subcapsular or perisplenic bleeding.

In the 3 cases in our series in which the splenic tip presented beneath the twelfth rib, pericapsular clots were found. The significance of this finding, however, becomes highly questionable when one realizes that it was present in 10 per cent of normal children. Further, the distance the splenic tip extended beneath the twelfth rib in the control cases was found to be equal or greater than that in the children with splenic injury. It is, therefore, more accurate to record splenic enlargement in 2 rather than 5 cases in Table 1. In the control series, where the splenic shadow could be identified, the cephalocaudal dimension varied from roentgenogram to roentgenogram with phase of respiration and projection.

### C. PLEURAL REACTION AT LEFT BASE

This was not found in any of our cases or in the controls. When present, it is nonspecific as any diaphragmatic irritation may produce it. It is, of course, important to realize that pleural reaction may not be due to an acute process but may represent the residue of antecedent supra- or infradiaphragmatic pathology.

### D. RIB FRACTURE

Although a frequently associated injury in adults, this was not found in any of our cases.

### E. BOWEL

1. *Widening of Space between Bowel Loops.* This did not occur in any of our cases or in the controls. This roentgen finding may be produced by fluid of any kind between loops of bowel or by any condition that thickens bowel wall.

2. *Distention of Both Large and Small Bowel.* This is attributed to reflex ileus and was present in one of our cases and in 9 controls. In the case of splenic fracture, this was due to diffuse intraperitoneal bleeding. In one of the controls it was due to acute appendicitis and in the others the cause was not determined. The younger the patient, the less is the significance of such bowel distention.

3. *Depressed Splenic Flexure.* This was found in one of our patients who had a large subcapsular hematoma. It was also found in 6 controls. In 3 it was secondary to splenomegaly, in 1 to a retroperitoneal tumor, in 1 to a renal mass, and in the last to a grossly distended stomach. It is apparent, then, that this finding is a function of consumption of left upper quadrant space.

### F. OBLITERATION OF VARIOUS SHADOWS

These signs are listed in Table 1. None was present in any of our cases. One or more of them was seen in 57 per cent of controls. In normal infants, visceral and psoas detail is poor and a ground glass haze is usually found. As the infant grows, perivisceral and parapsoas fat is deposited, making these structures more apparent but still difficult to see through overlying intestinal content. Thus, on serial roentgeno-

grams, as the intestines and/or their contents shift, varying segments of the psoas shadow become visible. Often the psoas shadow is seen only in an oblique view.

It was thought that roentgen findings other than those described in the literature might be of value. These are listed in Table II and are discussed below.

### ADDITIONAL ROENTGEN FINDINGS

#### A. INCREASE IN GASTROCOLIC SPACE

Since a large amount of any type of fluid in the lesser sac may increase this space, it is possible that blood from a ruptured spleen may do so. This measurement appeared markedly increased in 2 of our cases. In the controls, however, this distance varied to such a degree that no accurate basis for comparison could be established.

#### B. SCOLIOSIS TO RIGHT

Parapsoas irritation, as with perinephric abscess, can produce scoliosis to the opposite side. Since bleeding from the spleen can be retroperitoneal, a similar situation is possible. While present in 4 of our cases, this finding also occurred in 25 per cent of controls. Unfortunately, the pediatric age group is not known for ease of positioning, and apparent scoliosis is so frequent as to make this sign valueless.

#### C. SENTINEL DISTENTION OF TRANSVERSE COLON OR DISTENTION OF COLON TO SPLENIC FLEXURE

One of the authors (S.S.S.) has described colon distention to the splenic flexure in pancreatitis.[9] It was felt that this was due to localized spasm at the splenic flexure or to a reflex ileus of this portion of bowel. As blood is an irritant, this phenomenon might follow splenic fracture. Such distention was found in 1 of our cases and in 5 controls, the latter without adequate explanation. Segmental distention of the transverse colon is merely a form of sentinel loop. This occurred in 2 of our cases and in 4 controls, the latter again without adequate explanation. Both of these signs are nonspecific. The fact that they were seen in apparently normal children further detracts from their value.

#### D. OBLITERATION OF LEFT PROPERITONEAL FAT LINE

Appendicitis in the right lower quadrant and diverticulitis in the left lower quadrant may obliterate the adjacent portion of properitoneal fat. On the chance that blood in the left upper quadrant might obscure this segment of properitoneal fat, this finding was investigated. It was present in 2 of our cases. In 35 controls, the line could not be seen on either side. In only 72 controls could both lines be seen in their entirety.

With a child properly positioned, the midportion of both lines is normally not seen. Should the thoracic spine be bent to one side, the homolateral line is obliterated and the opposite is seen in its entirety. Further, as discussed under the section on psoas shadow, age, position and variation in bowel content affect the visibility of these lines. In view of the multiple factors influencing their visualization, the properitoneal fat lines may be seen in varying degree on serial roentgenograms of the same child.

### DISCUSSION

An analysis of the clinical and laboratory aspects of traumatic rupture of the spleen in 33 children has been presented in a previous paper.[2] The most significant fact determined in that study was the failure of two-thirds of the cases to show symptoms, signs or laboratory indications of blood loss. This is in contradistinction to the classic description of this condition which emphasizes the shock-like picture. The minimal clinical findings prompted this study in the hope of evolving a specific roentgen aid to diagnosis.

Although it is unfair to take individual criteria and demonstrate their fallibility, there appears to be no roentgen finding or group of findings which consistently add up to a diagnosis of splenic trauma. The most significant finding was exaggeration of mucosal folds on the greater curvature of the stomach, which was present in 7 of

our 12 cases. It also appeared, however, in 6 per cent of controls. The remaining signs, both those previously described in the literature and those investigated by us for possible value, occurred so infrequently as to make them of little importance. Further, the common demonstration of many of these so-called diagnostic signs on the abdominal roentgenograms of the normal children nullifies their significance. Moreover, the frequent inability to outline visceral and psoas shadows in this age group makes interpretation of alterations of these shadows impossible. Accessory aids such as thorotrast injection and pneumoperitoneum have been suggested.[1,3] The former should be comdemned especially in this age group and the latter seems to have no logical value.

Were the diagnosis of a ruptured spleen deferred because of the absence of the classically described clinical and/or roentgenographic findings, delay and failure in correct therapy would have resulted. The clinical picture which has evolved from our studies is completely different from that found in texts and in previously published series. The triad of: (1) history of trauma; (2) abdominal and/or shoulder pain; (3) abdominal tenderness and muscle guarding, especially when localized to the left upper quadrant, is in our cases far more typical of ruptured spleen in children than signs of blood loss. Early exploration of children presenting this picture regardless of roentgen findings will result in fewer catastrophies due to missed diagnosis and to an over-all lowering of morbidity.

SUMMARY

1. Abdominal roentgenograms of 12 children with traumatic rupture of the spleen have been reviewed.

2. Roentgenograms of 200 infants and children, most of whom were normal and the remainder suffering from conditions other than splenic rupture, were used as controls.

3. The only roentgen sign occurring with any degree of frequency in those cases with rupture of the spleen was exaggeration of the mucosal folds on the greater curvature of the stomach. This is not specific as it also occurred in 6 per cent of controls.

4. Previously published or other roentgen findings were either infrequent or not valid in this age group.

Solomon S. Schwartz, M.D.
Director of Radiology
The Jewish Hospital of Brooklyn
555 Prospect Place
Brooklyn 38, New York

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# IDIOPATHIC ADRENAL CALCIFICATION IN INFANTS AND CHILDREN*

*By* J. LUTHER JARVIS, M.D., *and* WILLIAM B. SEAMAN, M.D.
NEW YORK, NEW YORK

CALCIFICATION of the adrenal gland is usually an indication of a pathologic process. It has been found in association with tuberculosis, Addison's disease, Niemann-Pick's disease, cysts, adrenal hemorrhage and adrenal neoplasms, including carcinoma, neuroblastoma, pheochromocytoma and ganglioneuroma. There is also a group of patients with calcification in the adrenal gland who do not show evidence of adrenal disease. The purpose of this paper is to present a study of a group of infants and children with idiopathic adrenal calcification.

The group consists of 26 patients, 17 males and 9 females, with an age range from five days to twelve years (Table I). Calcification of the adrenal was diagnosed roentgenographically in 19, while 7 cases were obtained from the necropsy records of Babies Hospital. As necropsies were done subsequently on 3 of the 19 patients who died from causes unrelated to adrenal disease, a total of 10 patients was studied post mortem.

In all of the 19 patients, adrenal calcification was an incidental roentgenographic finding; indications for roentgen examinations had been varied, but unrelated to adrenal disease. Clinical follow-up (Table II) ranging from three to twenty years, obtained in 14 patients, showed no evidence of adrenal disease. In 2 of the remaining 5, the adrenal calcification had been detected only recently; the 3 other patients died shortly after identification of the calcification. Cutaneous tuberculin tests were negative in 16 patients. One patient (No. 17) with a positive tuberculin test was found at necropsy to have pulmonary, but not adrenal tuberculosis. In 2 patients the tuberculin test was not performed. Calcification was bilateral in 10 instances, only on the right side in 6, and only on the left side in 3.

TABLE I

AGE AT DETECTION OF ADRENAL CALCIFICATION

| Age | Living | Dead |
|---|---|---|
| 5–12 da. | | 2 |
| 1– 6 mo. | 1 | 4 |
| 7–12 mo. | 3 | 3 |
| 1– 3 yr. | 3 | 1 |
| 4– 6 yr. | 4 | |
| 7–12 yr. | 5 | |
| Total | 16 | 10 |

The calcified adrenal (Fig. 1*A*) appears roentgenographically as a collection of irregular, discrete densities which tend to be grouped in a triangular fashion. The large size of the calcified area in infants may seem surprising until one recalls that at birth the adrenal glands are relatively large bodies, about one-third the size of the kidneys. Immediately following birth, the adrenal cortex begins to involute. The glands decrease to two-thirds their birth weight during the first two weeks of life and to one-half their birth weight within four months.[22] In the very young infant, this involution may occasionally be reflected in the roentgenogram (Fig. 1*B*), when the area of calcification shrinks and becomes more opaque. A similar decrease in the size of the calcification may result at any age from shrinkage of fibrous tissue formed after hemorrhage. This condensing process was noted in another patient (Fig.

* From the Department of Radiology of the College of Physicians and Surgeons of Columbia University and the Columbia-Presbyterian Medical Center, New York, New York.

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TABLE II

IDIOPATHIC ADRENAL CALCIFICATION—LIVING PATIENTS

| Case No. | Age at Diagnosis (years) and Sex | Adrenal Calcification | Tuberculin Test | Follow-up (Length in years) | | Birth History | | | Associated Diagnoses |
|---|---|---|---|---|---|---|---|---|---|
| | | | | Clinical | Roentgenographic | Pregnancy and Delivery | Weight | Anoxia | |
| 1. D.T. (180616) | 8 F | Right | Neg. | 4 | 4 | Mid forceps extraction | 8 lb. | No | Mild hemiparesis at birth |
| 2. K.H. (041623) | 1 3/12 F | Left | Neg. | 8 | 6 | Long labor. Low forceps. Forceps scar on cheek 9 years after birth | 7 lb., 11 oz. | — | Cerebral atrophy by pneumoencephalography |
| 3. L.P. (002390) | 3/12 M | Bilat. | Neg. | 8 | 6 | Transverse lie with prolapse of umbilical cord and right arm. Podalic version and breech extraction. Resuscitation. Dislocation right shoulder. Temperature 108° 12 hours after delivery. Cyanosis | 6 lb., 11 oz. | Severe | Cerebral palsy |
| 4. R.C. (112825) | 3½ M | Bilat. | Neg. | 5½ | 3 | Forceps delivery. Partial facial paralysis. Forceps scar on forehead and right ear | 9 lb. | Yes | |
| 5. E.K. (122512) | 8 M | Right | Neg. | 6 | 5 | Kjelland rotation followed by mid forceps extraction | 7 lb., 2 oz. | No | Psychiatric problem |
| 6. N.N. (201118) | 3½ F | Bilat. | Neg. | 4 | 4 | Hydramnios. Induced labor. Low forceps. Maternal coccyx "cracked" as head came down | 10 lb., 8 oz. | No | Hydronephrosis and hydroureter |
| 7. P.W. (424906) | 5½ M | Right | — | 10 | 0 | No hospital confirmation | — | — | — |
| 8. A.A. | 1/12 M | Bilat. | — | 12 | 10 | Premature | 3 lb., 8 oz. | — | — |
| 9. W.C. (619740) | 11 M | Bilat. | Neg. | 3 | 0 | No hospital confirmation | 7 lb. | — | — |
| 10. P.M. | 3 M | Bilat. | Neg. | 8 | 8 | Spontaneous delivery, followed by cyanosis and hematuria | 9 lb., 8 oz. | Severe | — |
| 11. L.H. (744491) | 9 M | Bilat. | Neg. | 15 | 1 | Impacted shoulders. Resuscitation. Erb's palsy. Hematoma of left shoulder. Cyanosis | 9 lb., 7 oz. | Severe | Clinical diagnosis of cerebral hemorrhage |
| 12. E.C. (791171) | 10 F | Bilat. | Neg. | 8 | 3 | Contracted pelvis. 26-hour labor. Breech extraction | 7 lb., 9 oz. | No | — |
| 13. M.K. (508687) | 11/12 F | Right | Neg. | 20 | 20 | Rotation and mid forceps extraction | 10 lb., 9 oz. | No | — |
| 14. R.C. (1183201) | 4½ M | Right | Neg. | ½ | ½ | Low forceps | 8 lb., 12 oz. | No | — |
| 15. T.E. (229434) | 1/12 M | Right | Neg. | 3 | 2½ | Normal. Operation on right kidney | — | No | Multicystic right kidney |
| 16. | 2 F | Bilat. | Neg. | 0 | 0 | Fifteen hour labor. Breech delivery | — | — | Retarded speech |

Note: Case No. 8 is included by courtesy of Drs. H. Sidney Heersma and R. C. Hildreth, Kalamazoo, Michigan.
Case No. 10 is included by courtesy of Dr. J. M. Frawley, Fresno, California.
Case No. 16 is included by courtesy of Dr. H. G. Barber, Homer, Nebraska.

2, *A* and *B*) and apparently occurred in the case of Drucker and Rodriguez.[9] In 11 additional patients, roentgenograms made after intervals which varied from one to twenty years disclosed no change in the adrenal calcifications.

### PATHOLOGIC FINDINGS

Necropsies were performed in 10 cases (Table III); 4 of these patients had been examined roentgenographically during life (Fig. 3). The adrenal calcification was demonstrated in 3. Roentgen examination failed to detect the calcification in the fourth case, in which at necropsy the calcareous deposit was limited to the center of a microscopic cortical adenoma. (Cortical adenomas occur so frequently as to be considered a normal variant.) In 2 premature infants, not examined roentgenographically, the calcification was microscopic in size. In another case both adrenals were normal but calcification was found in an aberrant adrenal in the pelvis. None of these patients had clinical signs of adrenal insufficiency or evidence of tuber-

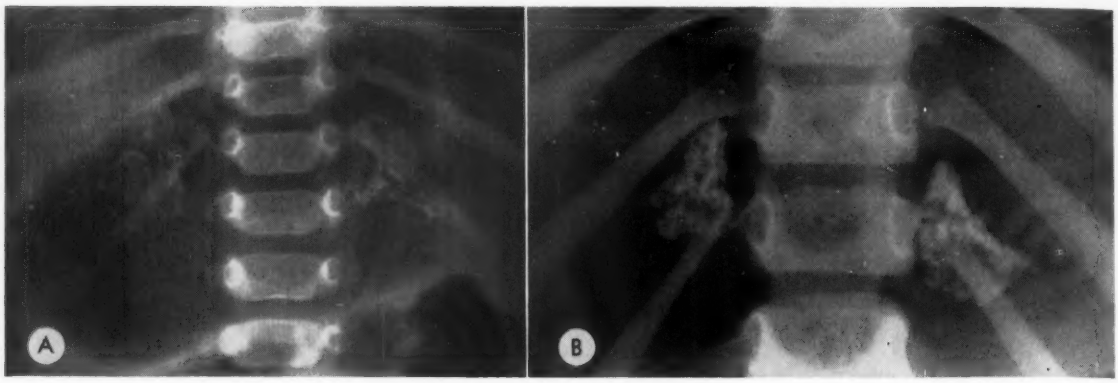

FIG. 1. Case No. 3, L.P. (*A*) June 27, 1950: Bilateral adrenal calcification in a two and one-half month old male infant who was delivered by internal podalic version and breech extraction, followed by intermittent cyanosis for two months. (*B*) May 18, 1956: Six years later the areas of calcification have diminished although the densities are greater. (Patient now has cerebral palsy.)

culosis with the exception of patient No. 17 (see above).

Microscopic examination usually showed replacement of the innermost portion of the cortex and medulla by calcareous deposits or by a mass of fibrous tissue containing calcium. The cortex was otherwise normal or slightly thinned in most cases in this series and in those reported in the literature (Table IV). The relative preservation of cortical structure may explain the absence of clinical signs of adrenal insufficiency. Deaths in this group were associated with sepsis in 5 patients, prematurity in 3 and brain damage at birth in 1. The cause of death was undetermined in 1. One patient, not included in this series, with calcification of the adrenal died of lupus erythematosus.

## DIFFERENTIAL DIAGNOSIS

Adrenal calcification must be distinguished from other calcifications in the same region. Calcification in gallstones, lymph nodes, costal cartilages and regional arteries may be confusing in the frontal projection but can usually be differentiated clearly in the lateral view. Calcification in the upper pole of the kidney may be more difficult to distinguish. Comparison of roentgenograms made with the subject in erect and supine positions may be helpful. According to Steinbach and Smith,[26] in the erect position the adrenal, which is more firmly attached to Gerota's fascia, drops less than the kidney; the greater drop of the kidney may separate it from the adrenal image.

Calcification in adrenal tumors presents

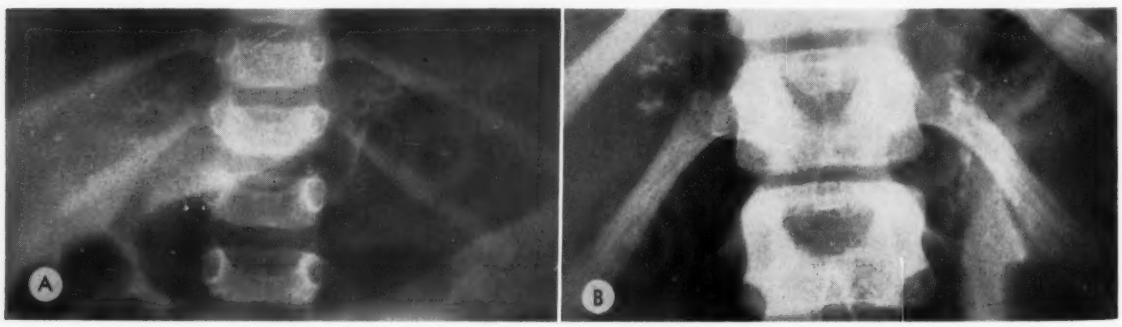

FIG. 2. Case No. 8, A.A. (*A*) Dec. 24, 1945: Bilateral adrenal calcification in an eight month old boy born prematurely. (*B*) May 13, 1956: More than ten years later, the calcifications are smaller, but denser. The patient is in excellent health with normal growth and development.

## Table III

IDIOPATHIC ADRENAL CALCIFICATION—NECROPSIED CASES FROM BABIES HOSPITAL

| Case No. | Age and Sex | Clinical Diagnosis | Tuberculin Test | Roentgen Findings | Birth History | | | Adrenal Calcification | | | Associated Diagnoses |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | Pregnancy and Delivery | Birth Weight | Anoxia | Location | Gross Calcification | Microscopic | |
| 17. J.O. (294599) | 8 mo. M | Suppurative pleurisy (hemolytic streptococcus) | Pos. | Bilat. | Breech. Difficult labor. Instrumentation. Dislocation left shoulder. Left 11th nerve palsy. Four cephalohematomas, still present 10 days after birth | 8 lb., 4 oz. | | Bilat. | Yes | Wide central zone containing large irregular calcific deposits. Also small irregular masses of calcium between cells of cortex. No tuberculosis | Tuberculosis of lung. Brain not examined |
| 18. G.F (308636) | 19 mo. M | Peritonitis, 1 week | Neg. | Left | Normal spontaneous delivery following 12 hour labor (excessive sized infant) | 11 lb., 12 oz. | | Left | Yes | Calcified mass occupying most of medulla. Small calcified masses in cortex | Brain not examined |
| 19. C.K. (673007) | 12 da. F | Premature | — | — | Premature twin. Normal spontaneous delivery. Cyanosis after birth. (Twin sibling: 1,550 gm. Facial twitchings began five hours after birth and ceased in a few hours, presumably central nervous system damage | (1,190 gm.) 2 lb., 10 oz. | Severe | — | No | Calcified thrombi beneath permanent cortex | Infarcts in kidney and spleen. Normal brain |
| 20. D.T. (677405) | 9 mo. M | Hyperpyrexia. Physical and mental retardation | Neg. | — | Breech. Head delivered 7 minutes after breech. Took 15 minutes to make infant cry. Hands clenched tightly into fists—did not open them for 6 weeks | 5 lb., 14 oz. | Yes | Bilat. | No | Medulla replaced by calcified mass. Cortex intact | Small hemorrhages of leptomeninges (clinical evidence of central nervous system damage) |
| 21. A.P. (689448) | 5 da. F | Premature | — | — | Premature 7 month twin (first delivered). Hydramnios. Normal spontaneous delivery at home | (1,300 gm.) 2 lb., 14 oz. | | Unilat. ? side | No | Incomplete involution of fetal cortex. A portion of remaining cortex shows calcium deposits of varying sizes | Subarachnoid hemorrhage, localized, right cerebral hemisphere |
| 22. I.L. (692830) | 2 mo. F | Congenital atresia of duodenum, operated on 7th day of life. Surgical complications | — | — | Premature. Hydramnios. Premature separation of placenta. Intermittent cyanosis for 5 days after birth | (1,550 gm.) 3 lb., 6 oz. | Severe | | | Normal adrenals. Calcification (central two-thirds) of aberrant adrenal in broad ligament. | Porencephaly in left and right parietal lobes, due to old hemorrhage |
| 23. R.F. (765130) | 6 mo. M | Meningitis and brain abscess | — | — | Premature 7½ month delivery. Frank breech extraction | (1,970 gm.) 4 lb., 6 oz. | | Unilat. ? side | No | Cortex and medulla normally developed. Irregular deposition of calcium between medulla and cortex | Meningitis (H. influenzae). Internal hydrocephalus, secondary |
| 24. J.H. (853604) | 4 mo. M | Hydrocephalus, primary | — | — | Breech. Forceps used after 27 hours of labor. Rapid breathing. Scalp lacerations. Hematomas in the left neck and mastoid region | Unknown | Yes | Unilat. ? side | No | Cortex thin and normal. Irregular deposits of calcium between reticular zone and medulla | Old hemorrhage in roof of fourth ventricle |
| 25. T.M. (032552) | 3½ mo. M | Brain abscess. Retrolentil fibroplasia | — | Neg. | Premature. Normal spontaneous delivery followed by cyanosis | (1,530 gm.) 3 lb., 6 oz. | Severe | Unilat. ? side | No | Cortical adenoma showing central fibrous area, which contains calcium | Cerebral abscess. Retrolental fibroplasia |
| 26. W.S. (187726) | 10 mo. M | Bladder neck obstruction. Abscess of kidneys | Neg. | Left | Normal spontaneous delivery | 8 lb., 11 oz. | | Left | Yes | Cortex normal. Large fibrosed and heavily calcified area replacing medulla | Encephalomalacia (old), cystic, of left frontal lobe |

## TABLE IV

### IDIOPATHIC ADRENAL CALCIFICATION—REPORTED IN THE LITERATURE

| Source | Age and Sex | Clinical Diagnosis | Roentgen Findings | Birth History | | Adrenal Calcification | | | Associated Diagnoses |
|---|---|---|---|---|---|---|---|---|---|
| | | | | Pregnancy, Delivery and Birth Weight | Anoxia | Location | Gross Calcification | Microscopic | |
| Snelling and Erb[23] | 5½ mo. M | Mastoiditis | | Breech. Difficult delivery. Hematoma of buttocks. Depressed fracture. Difficult resuscitation. High rasping cry for 2 weeks | Yes | Bilat. | No | Numerous areas of calcareous deposits bilaterally | |
| | 4¾ mo. M | Peritonitis | | Normal labor. 8 lb. | Unknown | Right | Yes | Cortex normal. Medulla entirely replaced by calcareous deposits and fibrous tissue | |
| | 9½ mo. M | Bronchopneumonia | Bilat. | Not stated | Unknown | Bilat. | Right | Left: veins thrombosed in medulla with some calcification. Right: many calcareous deposits. Medulla all destroyed. Cortex only a thin shell | |
| | 7 mo. F | Otitis media | Bilat. | Normal labor | Unknown | Bilat. | Yes | Medulla and part of cortex replaced by calcareous deposits. In one there was formation of osseous tissue with bone marrow | |
| | 17 mo. M | Convulsions | | Not stated | Unknown | Bilat. | Yes | Cortical cells normal, but structure altered by large old medullary hemorrhage containing calcification | |
| | 7 da. M | Hemorrhage | | Breech delivery. Forceps. Difficult resuscitation. Irregular shallow breathing. Did not move right side of body. Mass appeared under the chin. Mass felt in abdomen | Yes | Right | No | Right: marked diffuse hemorrhage with calcareous deposits. Left: completely destroyed and replaced by blood clot | Intracranial hemorrhage, subarachnoid |
| | 6 da. F | Intra-abdominal hemorrhage and cerebral hemorrhage | | Prolonged instrumental labor | Yes | Bilat. | No | Thin shell of cortex (in both glands) surrounding blood clot. Patches of calcareous deposit immediately under cortex | Retroperitoneal hemorrhage. Cerebral hemorrhage clinically |
| | 5½ yr. M | Midline cerebellar tumor | | Not stated | Unknown | Right | Yes | Adrenal fibrosed with very little gland tissue remaining. In the center of the fibrous tissue were calcareous deposits and bone formation with marrow spaces | Tumor of hypothalamus |
| Drucker and Rodriguez[9] | 21 mo. F | Convulsions (negative tuberculin test) | Bilat. | Premature. Frank breech. Forceps used on after-coming head. Slow tremors of mouth and lower extremities. Neurosurgical evaluation: brain damage. 5 lb., 1 oz.[17] | Unknown | Bilat. | Yes | Pronounced calcification with necrosis of the medulla and a portion of the adjacent reticular zone of the cortex | |
| Stevens and Tomsykoski[27] | 4 wk. F | | | Breech. Protracted and difficult delivery. Right brachial palsy. Hands and feet blue. 7 lb., 4 oz.[17] | Yes | Bilat. | No | Large deposits of calcareous material between central necrotic area and a narrow zone of normal cortical cells | |
| Rack and Eiben[21] | 22 mo. F | ? Neuroblastoma<br>? Neuroblasts on marrow smear<br>Right adrenal calcification seen on skeletal survey. (Beaten by father 6 mo. before, causing right flank pain.) Right adrenalectomy | | Normal pregnancy with delivery at term, vertex presentation with forceps[17] | No | Right | Yes | Normal architecture distorted by large areas of old and recent hemorrhage containing calcific plaques, many arranged concentrically | |
| Williams[30] | 3 mo. M | ? Neuroblastoma<br>(Meningitis and urinary tract infection on admission.) Large calcification displacing kidney seen on retrograde pyelogram. Left adrenalectomy | | Surgical induction. Normal labor. Weight 10 lb.[17] | Unknown | Left | Yes | Normal architecture entirely destroyed. Small isolated islets of cortical tissue remained among masses of acellular tissue and looser vascular tissue containing extensive calcification | |
| Lintz[13] | 11 yr. M | Epilepsy | | Not stated | Unknown | Bilat. | | | 6 yr. clinical follow-up |
| Potter[19] | 9 mo. | | | Not available[17] | | | | Organized and partially calcified hemorrhage in center of adrenal gland | |
| Nicter[20] | 14 mo. M | Sudden death | | Breech delivery. Marked asphyxia. Re- | Yes | Bilat. | Yes | Left: medulla surrounded by fibrous tis- | |

a more difficult diagnostic problem. When calcification is bilateral, the possibility of a neoplasm is small, but when it is unilateral, as was the case in 9 patients of our group, neoplasm cannot be excluded conclusively. The neoplastic mass may not be visible on conventional roentgenograms of the abdomen and special procedures, such as retroperitoneal pneumography, may be necessary for its detection.

Neuroblastoma is the most frequently calcified abdominal tumor of infancy and childhood. Calcification is roentgenographically demonstrable in about 50 per cent of neuroblastomas but in only 8–12 per cent of Wilms' tumors.[14] Thus, when adrenal calcification is associated with a mass, neuroblastoma is the most likely possibility. Conversely, the absence of a mass supports the diagnosis of idiopathic adrenal calcification (Fig. 4, *A* and *B*). However, some neuroblastomas are very small (Fig. 5), while a calcified adrenal associated with a large hematoma, may roentgenographically simulate a neoplasm. Two patients reported in the literature had adrenalectomies because of clinical diagnoses of neuroblastoma.[21,30] One of these, described by Williams,[30] was an infant with a calcified left adrenal mass which displaced the kidney. At operation this proved to be a large, partially calcified hematoma within the adrenal.

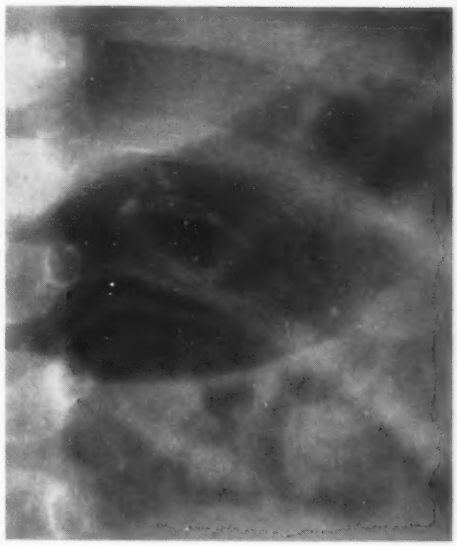

Fig. 3. Case No. 26, W.S. Unilateral adrenal calcification in a ten month old male who died of generalized sepsis associated with bladder neck obstruction. At necropsy the adrenal cortex was normal, but the medulla was replaced by fibrous tissue and extensive calcification.

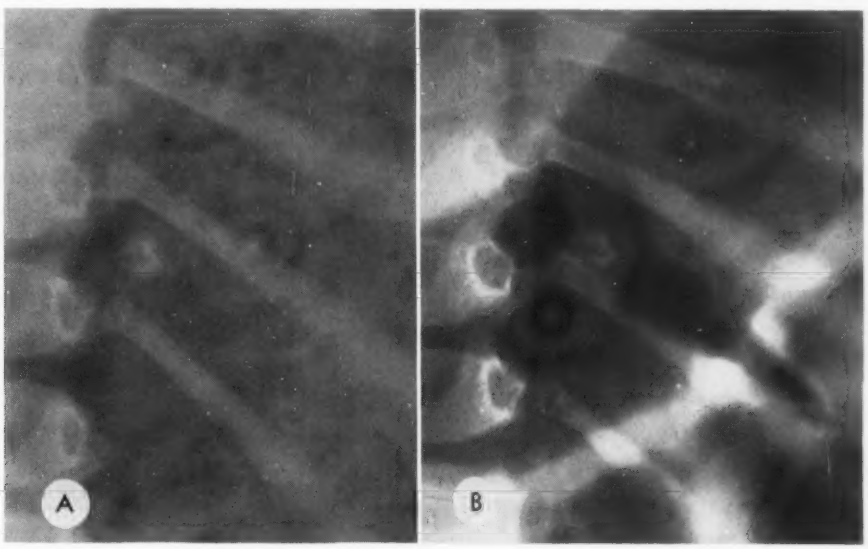


Fig. 4. Case No. 2, K.H. (*A*) Unilateral adrenal calcification in a fourteen month old girl delivered by low forceps after a long labor. Pneumoencephalography revealed dilated ventricles and widened sulci compatible with cerebral and cortical atrophy. (*B*) Roentgenogram of the left adrenal gland after presacral air injection shows no mass or adrenal enlargement.

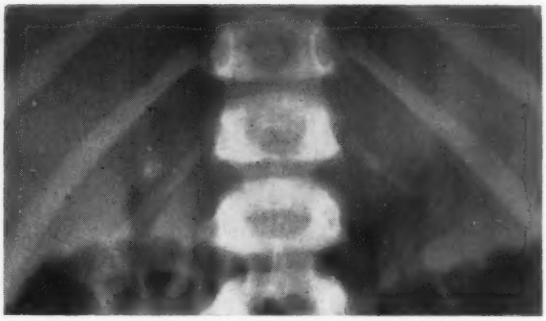

FIG. 5. Calcification in the right adrenal area of a patient with multiple osseous metastases from a neuroblastoma (histologically confirmed). No mass is visible in this roentgenogram.

Less common causes of adrenal calcification are adrenocortical carcinomas,[14] pheochromocytomas,[24] adrenal cysts,[3] Niemann-Pick disease,[1,2,6] and Addison's disease.[29] Adrenocortical carcinomas are frequently associated with either Cushing's disease or the adrenogenital syndrome. Pheochromocytomas are relatively uncommon in children. Three cases of adrenal calcification in Niemann-Pick disease have been reported; in all 3 the calcification was massive and limited to the medulla leaving a thin margin of cortex. Addison's disease is also rare in infants and children, but 4 cases were recently reported by Williams and Robinson[29] who described fibrosis, hemosiderin deposits and foci of calcification in the adrenals of two of their patients. Thus it is possible that in some instances adrenal hemorrhage and subsequent calcification may be associated with adrenal insufficiency.

## PATHOGENESIS

It has been noted by many observers that adrenal hemorrhage in the newborn is not necessarily fatal but may become calcified or even ossified.[16,23] In a series of 3,637 consecutive necropsies in infants and children, Snelling and Erb[23] found 43 examples of adrenal hemorrhage, 8 of which were associated with calcification. The mechanism of the adrenal hemorrhage has never been satisfactorily explained, although it is known that bleeding begins in the central part of the gland.[19] Potter[19] states that it is most often seen in infants delivered from a breech position, presumably a result of trauma sustained by the fetal abdomen during delivery of the head. The relationship of trauma to adrenal hemorrhage and subsequent calcification is well illustrated by the following case:

This infant (No. 15) was explored surgically at eight days of age for a right-sided abdominal mass. A large kidney which contained multiple cysts was found; a biopsy specimen was not taken. Preoperative roentgenogrmas (Fig. 6 *A*) of the gastrointestinal tract revealed no adrenal calcification. The postoperative course and subsequent development were normal. However, adrenal calcification was demonstrable eight months later (Fig. 6 *B*) and again at the age of eighteen months when a retrograde pyelogram was made (Fig. 6 *C*). At thirty-six months the child was still asymptomatic and the adrenal calcification was unchanged.

In another patient (No. 22), the possible sequence of direct trauma, adrenal hemorrhage and subsequent calcification must be considered. A premature infant was operated upon seven days after birth for congenital atresia of the duodenum. At necropsy seven weeks later both adrenal glands were normal, but an aberrant adrenal gland with calcification in the central two-thirds was found in the broad ligament. During examination of the lower abdomen, the surgeon may have inadvertently traumatized the aberrant adrenal gland or compromised the blood supply.

## BIRTH TRAUMA

Parturition is a potential source of trauma for infants; therefore, the birth histories of the 26 infants and children were carefully studied (Table v). Twenty-four were delivered in 23 different hospitals, one was born at home, and for one the obstetric history could not be obtained. In a number of instances, the birth history was recorded as normal on the patient's hospital chart, but when an accurate obstetric record was obtained, the birth was found to have been abnormal.

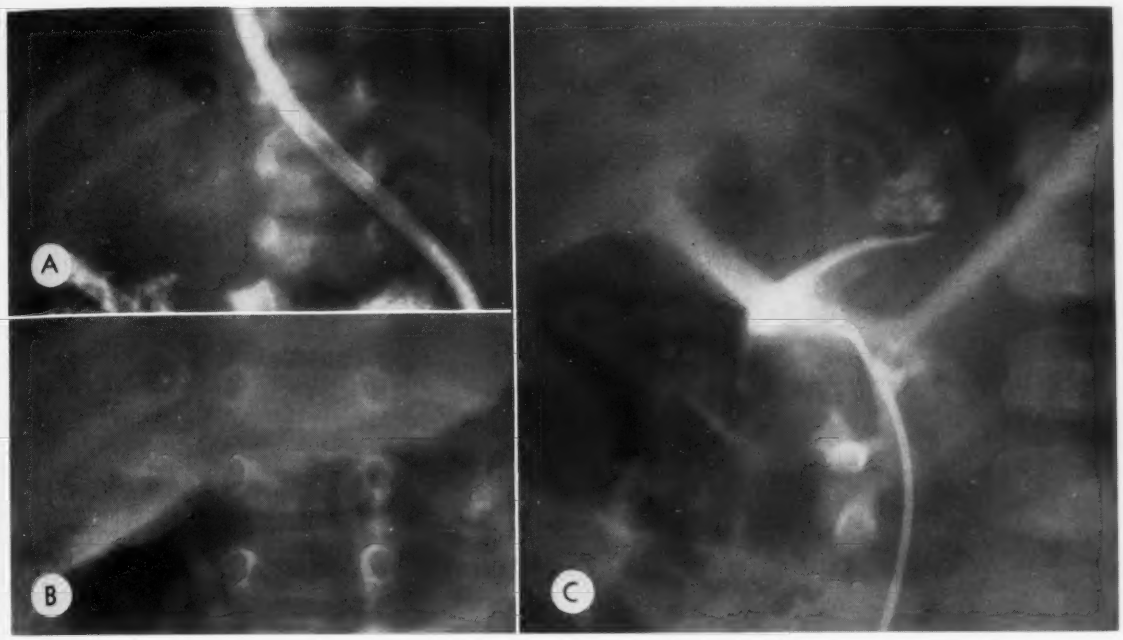


FIG. 6. Case No. 15, T.E. (*A*) May 24, 1955: Preoperative roentgenogram at eight days of age shows no adrenal calcification. (*B*) Jan. 19, 1956: Eight months later the right adrenal area is calcified. (*C*) Nov. 7, 1956: Retrograde pyelogram taken eighteen months later. Inadvertent subcapsular injection of contrast medium clearly demonstrates calcification outside of the renal capsule and in the region of the adrenal gland.

A review of the obstetric histories of the 16 living patients revealed that 13 had had abnormal births. In 2 instances detailed birth information could not be obtained; in another (No. 15) adrenal calcification developed following renal surgery. Seven patients were delivered by forceps, 3 had breech deliveries, one premature infant weighed 3½ pounds and 2 were "excessive sized" infants (2 additional oversized infants are discussed with the group delivered by forceps).

Mid-forceps extractions occurred in 3 instances. These babies weighed 8 pounds; 7 pounds, 2 ounces; and 10 pounds, 9 ounces. Low forceps was applied in the case of a large infant weighing 10 pounds, 8 ounces, in another baby who weighed 8 pounds, 12 ounces, and in a third birth after prolonged labor. A 9 pound infant delivered by forceps had permanent scars on the face and later developed facial paralysis. Of the 3 breech deliveries, one was by breech extraction after twenty-six hour labor, another by version and breech extraction because of a transverse lie with prolapsed cord and arm, and the third by a breech extraction of an unknown type following fifteen hour labor.

"Excessive sized infant" is a term used by Eastman[10] (and at Sloane Hospital of the Columbia-Presbyterian Medical Center) to describe any baby weighing over 9 pounds (4,000 gm.) at birth; Gollin *et al.*[11] use it to designate a newborn weighing 10

TABLE V

OBSTETRIC HISTORY

| Delivery | Living | Dead | Total |
|---|---|---|---|
| Forceps | 7 | | 7 |
| Breech | 3 | 3 | 6 |
| Premature | 1 | 5 | 6 |
| Birth injury | 1 | | 1 |
| Excessive size | 1 | 1 | 2 |
| Normal | 1* | 1 | 2 |
| Insufficient history | 2 | | 2 |
| Total | 16 | 10 | 26 |

* Renal surgery.

pounds (4,500 gm.) or over. The latter reported that, in 40,944 deliveries, infants weighing 10 pounds or more represented only 0.488 per cent of the total deliveries. However, the antenatal death rate was 7.7 times greater in the excessive sized infant and the intranatal death rate 15.6 times greater, presumably due to difficult delivery.

Four of the 16 living patients weighed over 9 pounds at birth. Two infants (weighing 10½ pounds and 10 pounds, 9 ounces) were delivered by forceps and are included with the forceps deliveries listed in Table v. Another large infant (9 pounds, 8 ounces) was cyanotic for the first five days of life with associated respiratory difficulties and hematuria. The fourth (9 pounds, 7 ounces) was born with Erb's palsy following impacted shoulders during delivery; this case is listed in Table v as a birth injury.

Examination of the birth histories in the 10 cases examined post mortem disclosed 5 premature infants (No. 19 and No. 21, from two sets of twins), 2 instrumental breech extractions, 1 breech delivery of unknown type and 1 excessive sized infant weighing 11 pounds, 12 ounces. One baby, who weighed 8 pounds, 11 ounces, had a normal birth history but died at ten months from uremia and overwhelming sepsis. In the 15 cases found in the literature, there were 4 breech deliveries of unknown type, 2 forceps deliveries of unknown type, 1 excessive sized infant weighing 10 pounds, 1 premature infant and 7 with insufficient birth information (Table iv).

Thus, 22 of 26 patients had abnormal obstetric histories, including cases of forceps delivery, prematurity, breech delivery and excessive sized infants. In forceps deliveries higher application increases the possibility of serious injury to the infant; an infant mortality of 3 to 10 per cent has been reported in several series of cases.[4,7,8,18,25] In breech deliveries survival rates are much lower than in vertex cases; the mortality rate of breech delivery is around 10 per cent.[20] Infants with birth weights of 6 to 8 pounds have the best chance for survival; the hazards of birth are increased for those whose birth weight is below or above these limits.[5] Premature infants are more susceptible to all conditions that adversely affect the more mature infant. The mortality rate for premature infants is 22 times that for infants born at term.[19]

### ANOXIA

Anoxia, according to Nesbitt,[15] may be a factor in the production of adrenal hemorrhage. The incidence of anoxia in premature infants is 12 times that for infants weighing over 2,500 gm.[19] In the group of living patients, 4 of 13 showed clinical evidence of anoxia at birth; 3 of these had intermittent cyanosis for several days. Five of the 10 examined at necropsy had been anoxic, including 3 cyanotic premature infants.

### ABNORMALITIES OF THE CENTRAL NERVOUS SYSTEM

An increased incidence of head trauma has been noted by Potter[19] in babies delivered by version and extraction, by mid and high forceps and from a breech position. Prolonged labor also appears to increase the risk of intracranial injury, according to Keith and Norval.[12] However, gross intracranial hemorrhage is occasionally seen after apparently normal, noninstrumental deliveries.[12] The incidence of intracranial hemorrhage among premature infants is five times that for infants delivered at term.[19]

Three of the living patients initially showed evidence of brain damage including mild transient hemiparesis and cerebral atrophy proved by pneumoencephalography; a fourth now has cerebral palsy. Four of 8 patients showed abnormalities in the central nervous system at post mortem (brain not examined in 2); in an additional case there was clinical evidence of intracranial damage. The obstetric histories of these 9 infants are recorded in Table vi; in 8 the births were abnormal.

Anoxia has been observed in association with central nervous system damage.[19] Two living patients in this study, who had evidence of central nervous system damage, including the child who now has cerebral palsy, were cyanotic at birth. Two patients, anoxic at birth, showed brain damage at necropsy. Thus, anoxia may be a causal factor in the production of abnormalities in the central nervous system as well as calcification in the adrenals.

TABLE VI

OBSTETRIC HISTORIES IN 9 INSTANCES OF CENTRAL NERVOUS SYSTEM ABNORMALITIES AND ADRENAL CALCIFICATION

| | |
|---|---|
| Premature | 2 |
| Breech extraction (instrumental) | 1 |
| Podalic version and breech extraction (for transverse lie and prolapsed cord) | 1 |
| Breech (unknown type) | 1 |
| Mid forceps extraction | 1 |
| Shoulder impaction (infant born with Erb's palsy) | 1 |
| Low forceps (for long labor) | 1 |
| Normal | 1 |
| Total | 9 |

## SUMMARY

1. A series of 26 patients with idiopathic adrenal calcification is presented. The calcification was demonstrated roentgenographically in 19 patients; in 7, it was discovered at necropsy.

2. Fourteen of 16 living patients, initially asymptomatic, have been followed from three to twenty years and continue to be free of disease.

3. The differential diagnosis of adrenal calcification is discussed.

4. Abnormal birth histories were found in 22 patients. There was a significant association of abnormalities in the central nervous system with birth trauma and anoxia.

5. Birth trauma and anoxia are believed to be important in the production of adrenal calcification.

J. Luther Jarvis, M.D.
Columbia-Presbyterian Medical Center
622 West 168th Street
New York 32, New York

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VOL. 82, NO. 3

# VOIDING CYSTOURETHROGRAPHY IN THE DIAGNOSIS OF CONGENITAL POSTERIOR URETHRAL VALVES

*By* WALTER A. GRIESBACH, M.D.,* REGINALD K. WATERHOUSE, M.D.,† *and* HARRY Z. MELLINS, M.D.‡
BROOKLYN, NEW YORK

CONGENITAL malformation of the posterior urethra, producing valvular obstruction to the urinary outflow and serious renal damage, constitutes one of the most urgent urologic problems of infancy and childhood. Nevertheless, statistical data indicate a decided variance in the accuracy of diagnosis of this condition from clinic to clinic. This suggests that not infrequently the correct diagnosis is arrived at late or not at all. The fact that cure rates have not risen in proportion to improvements in therapeutic techniques lends support to this conclusion. The key to the problem appears to lie in the method employed in the study of these patients. The technique of voiding cystourethrography is of proved superiority to retrograde urethrography and cystoscopy. The diagnostic features of congenital posterior urethral valves and the advantages of the voiding cystourethrogram are illustrated in the cases described in this report.

## PATHOLOGY

Congenital posterior urethral valves were first described over one hundred and fifty years ago. In 1919 Young, Frontz and Baldwin[21] reported the first preoperatively diagnosed cases and described three types of valves. Type I, inverted "V" shaped folds projecting distally from the verumontanum, is commonly recognized. Agreement on the existence of Type II, similar folds extending proximally from the verumontanum, and Type III, the perforated diaphragmatic type of obstruction, is not universal although these are described in the literature. Little has been added to the knowledge of the etiology of these valves and no single theory of their origin satisfactorily explains all cases. Although instances of urethral valves have been described in females, this is essentially a disease of the male child.

Among the causes of lower urinary tract obstruction, besides congenital valves, are meatal stenosis, neurogenic bladder and primary bladder neck obstruction. It is important to recognize that bladder neck obstruction may also occur as a result of or in conjunction with one of the other obstructive phenomena. Failure to properly evaluate this leads to improper management and therapeutic failure as demonstrated by one of the cases (Case II) described below.

## CLINICAL MANIFESTATIONS

The signs and symptoms of posterior urethral valves are those of any obstruction to the lower urinary tract in children. These are bladder distention, nocturnal or diurnal enuresis, frequency and urgency, urinary tract infection, disturbances of growth and development, hematuria, and abnormality of urinary stream. They are often mild and may not be recognized by parents who have never seen their child void normally. Difficulty in evaluation of these signs and symptoms arises from the fact that they closely simulate those of emotional disturbance. Any of the above manifestations should lead to examination of the lower and upper urinary tracts. Azotemia and renal insufficiency are late manifestations and their appearance indicates a poor prognosis.

* Instructor, Department of Radiology, State University of New York Downstate Medical Center, Brooklyn, New York.
† Instructor, Department of Urology, State University of New York Downstate Medical Center, Brooklyn, New York.
‡ Professor and Chairman, Department of Radiology, State University of New York Downstate Medical Center, Brooklyn, New York.

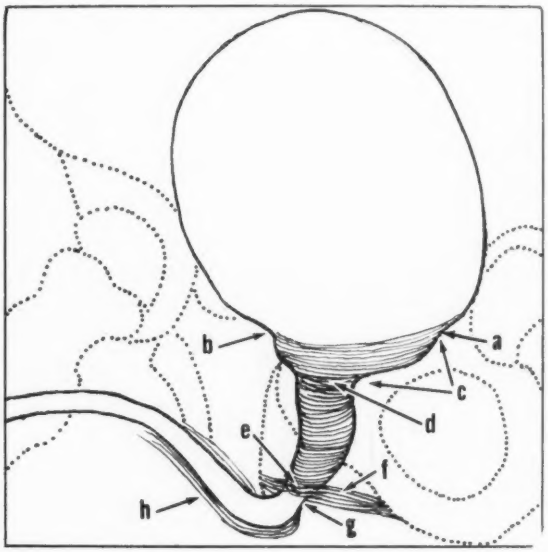


FIG. 1. The posterior urethra. The shaded portions represent detrusor muscle, voluntary muscle of the urethra, and deep transverse muscle fibers from the urogenital diaphragm (a, interureteric ridge; b, fundus ring; c, trigone; d, internal sphincter; e, external sphincter; f, urogenital diaphragm [deep transverse muscle]; g, pars nuda [membranous urethra]; h, bulbocavernosus muscle).

## NORMAL ROENTGENOGRAPHIC ANATOMY

For the evaluation of pathologic changes, a thorough familiarity with the normal roentgen anatomy of the urethra on the voiding cystourethrogram is necessary. Physiologic variations in contractions of the trigonal region of the urinary bladder and in the sphincter mechanisms of the urethra during micturition are responsible for the dissimilarity in the appearance of the posterior urethra on instantaneous roentgenograms. These factors make roentgenographic evaluation of the bladder neck region particularly difficult. Cinefluorography may shed further light on the normal anatomy of this area.

The upper extent of the urethra, the internal sphincter, is produced by interdigitation of fibers of the detrusor muscle as they extend downward over the junction of the urethra and the bladder (Fig. 1). Below this the urethra is surrounded on three sides by the prostate. The verumontanum bulges into the dorsal wall of the urethra approximately midway between the internal and external sphincters. The posterior urethra is surrounded by a coat of voluntary muscle fibers which act to produce contraction of the urethra. These interdigitate with muscle fibers from the urogenital diaphragm to form the lower extent of the prostatic urethra, the external sphincter. As the urethra pierces the urogenital diaphragm, there is a deficiency of muscle between the external sphincter and the origin of the bulbocavernosus muscle of the anterior urethra, the pars nuda or membranous urethra.

These anatomically separated portions of the posterior urethra may or may not be distinguishable on the roentgenogram depending on the physiologic state at the time the film is exposed. Figures 2 to 7 are examples of normal voiding cystourethrograms demonstrating some of the physiologic variations. Failure of relaxation of the internal sphincter has been described by Kjellberg, Ericsson and Rudhe[9] in children with and without urinary disturbances. Figures 3 and 4 demonstrate this phenomenon as a temporary manifestation in a

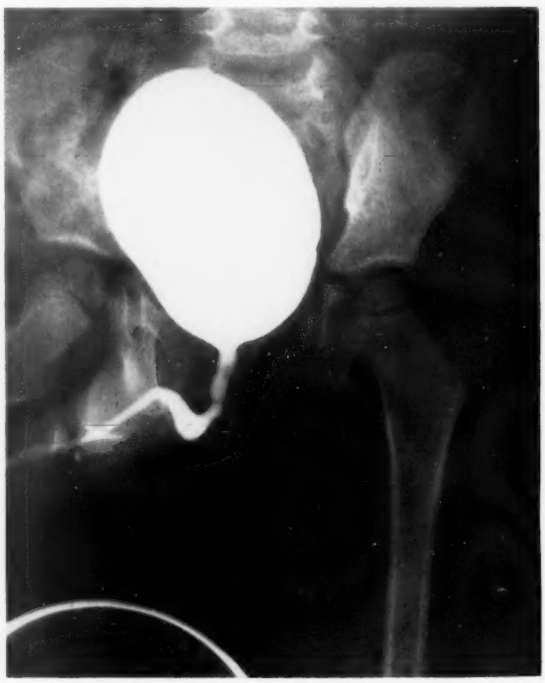

FIG. 2. Normal voiding cystourethrogram.

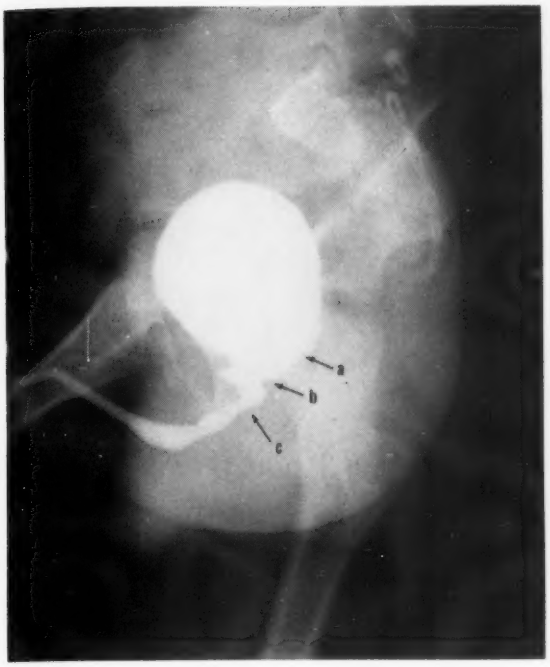


Fig. 3. Normal voiding cystourethrogram (a, interureteric ridge; b, contracted trigonal area called by Kjellberg *et al.* "fundus ring"; c, internal sphincter).

Fig. 4. Same case as in Figure 3, a few moments later.

Fig. 5. Normal voiding cystourethrogram. Nonobstructive folds are evident.

child examined because of nocturnal enuresis.

Nonobstructive folds have been described by Williams[19] and by Kjellberg *et al.*[9] These produce filling defects similar in appearance to that of valves, but do not cause dilatation of the proximal urethra (Fig. 5). They may represent mucosal folds or a transitional state between these and true valves. We encountered a rather striking

Fig. 6. Normal voiding cystourethrogram (after intravenous pyelography). Note again a contracted "fundus ring." This is probably a normal temporary phenomenon during micturition.

FIG. 7. Normal voiding cystourethrogram. The urethra is distended to the upper limits of normal.

example of this condition during the evaluation of a five year old child for persistent nocturnal and diurnal enuresis. The first study (Fig. 8) showed bladder trabeculation and questionable widening of the posterior urethra. The findings were confirmed

FIG. 8. The posterior urethra appears questionably dilated.

FIG. 9. Same case as Figure 8. A filling defect (arrow) is suggestive of valves. The posterior urethra appears somewhat more dilated.

by cystoscopic examination. No abnormalities of the urethra were seen; however, narrowing of the bladder neck was thought to account for the findings. A second voiding cystourethrogram (Fig. 9) suggested valves, but none could be seen during repeat cystoscopic examination. The upper urinary tracts were normal. Surgical exploration of the urethra and bladder neck was proposed but was not permitted by the parents. To date the symptoms have persisted, but nothing further has been done. A voiding cystourethrogram five months later was identical in appearance to that seen in Figure 9. The exact nature of these changes remains obscure. It appears highly unlikely that these represent true congenital valves in view of the lack of changes in the upper urinary tracts. Prolonged follow-up may provide an answer to this problem.

## TECHNIQUE OF VOIDING CYSTOURETHROGRAPHY

We have used the following method for obtaining voiding cystourethrograms:

1. The patient is asked to empty the bladder if possible. The urinary stream should be observed at this time.
2. With the patient supine a urethral

catheter is passed using sterile technique. Note is taken of any residual urine.

3. A 20 to 30 per cent solution of any urographic contrast medium is injected into the bladder. A total of 75 to 200 cc. is used, depending on the size of the patient and signs and symptoms of urge to void.

4. The patient is rotated toward the right 60 degrees or more, with the right thigh flexed and the left extended. The catheter is removed and an exposure is made during micturition. If the patient is able to cooperate, a second oblique roentgenogram as well as a straight anteroposterior roentgenogram should be obtained.

We have been successful in performing this examination even in the newborn. Figures 10 and 11 show normal urethrograms in three day and twelve day old infants. For such cases it is necessary, prior to removal of the catheter, to have everything prepared for a single exposure. At times, much patience is required and an alert technician must expose the film during the few seconds that a steady urinary stream is maintained. Older children may find it considerably easier to void on request in a standing position. If the roentgenographic equipment permits an upright roentgenogram, this may be obtained in the right posterior oblique position with the patient's right thigh elevated by placing the foot on a footstool. In all cases, since ureteral reflux often occurs only during micturition, a film large enough to include the upper urinary tract should be used. Examination of the upper urinary tracts by excretory urography must be postponed if reflux has occurred. Otherwise, it may be performed immediately following the voiding cystourethrogram.

Fig. 11. Normal voiding cystourethrogram in a twelve day old infant.

Fig. 10. Normal voiding cystourethrogram in a three day old infant.

### Report of Cases

Case 1. Baby boy M., three days old, was referred to the urologic service because of paraphimosis and a transilluminable scrotal mass. Initial physical examination was within normal limits but later there was intermittent abdom-

FIG. 12. Case I. Note filling defect suggesting valves below the dilated posterior urethra and the constriction at the base of the bladder.

inal distention. A left flank mass and a midline suprapubic mass were palpated at this time. The latter became smaller after rather difficult catheterization. Spontaneous micturition "with a good forceful stream" was recorded in the nursing notes. Excretory urography was not satisfactory but did suggest bilateral function. A cystogram revealed "bladder trabeculation and possible dilatation of the internal sphincter" (Fig. 12). Nevertheless, in the urologist's opinion the urinary tract was within normal limits. Medication included penicillin, chloromycetin and gantrisin. One month after birth, twitchings of the extremities were reported. Respiratory difficulties and cardiac arrhythmias appeared periodically and at the age of thirty-five days the infant expired. At necropsy obstructing posterior urethral valves were found with bilateral hydroureter and hydronephorsis.

*Comment.* In retrospect the diagnosis could have been suggested on the basis of the cystogram which demonstrates a dilated posterior urethra. On reviewing the roentgenograms we believe that the appearance was pathognomonic for urethral valves.

CASE II. J.S., a poorly developed seven year old Negro boy, was admitted to the Kings County Hospital with complaints of urinary incontinence, urgency, frequency and difficulty in starting micturition. A "Y" cystoplasty was performed at another hospital at the age of two years for similar complaints. Urinalysis and chemical analysis of the blood were normal. Excretory and retrograde urography showed bilateral delayed function, hydronephrosis and hydroureters. Cystoscopic examination revealed a "narrowed bladder neck." A voiding cystourethrogram (Fig. 13) demonstrated a dilated posterior urethra. This was not appreciated by the urologist and a second "Y" cystoplasty was performed on the same day. The postoperative course was uneventful. On repeat cystoscopic examination posterior urethral valves were seen. Excretory urography at this time showed no evidence of function on the right side and increased hydronephrosis on the left side. Three days later the third surgical procedure, removal of the valves, was undertaken. Since that time the patient's upper urinary tracts have not changed significantly, although the appearance of the urethra is now improved. There has been no return of function on the right and no improvement of

FIG. 13. Case II. The posterior urethra is dilated; valves are not clearly demonstrated. The deficiency in the region of the bladder neck (arrow) is presumably due to bladder neck obstruction similar to that seen in Figure 12.

the hydronephrotic changes on the left. The child has had repeated bouts of urinary infection. It is felt that the marked dilatation and kinking of the ureters still hinder adequate urinary drainage. The prognosis is guarded and further management will depend on the clinical course.

*Comment.* It seems worthwhile to emphasize that the correct diagnosis was made on cystoscopic examination only after the voiding cystourethrogram demonstrated the lesion.

Case III. K.B., a twenty month old Negro boy, was admitted because of a lower abdominal mass. History revealed that the child had ceased to develop at twelve months and had had recurrent abdominal swelling with fever, polyuria and polydipsia. On physical examination he appeared chronically ill. The abdomen was protuberant. A suprapubic mass and a left flank mass were noted. Urinalysis and chemical analysis of the blood were normal. The lower abdominal mass shrank following micturition.

Excretory urography showed nonfunction on the right and delayed function with marked hydronephrosis on the left side. Retrograde pyelography revealed bilateral hydronephrosis and hydroureter. The voiding cystourethrogram showed dilatation of the posterior urethra, bladder diverticula and ureteral reflux (Fig. 14). The presence of valves, postulated on the basis of the roentgenographic appearance, was confirmed on cystoscopic examination.

Fig. 14. Case III. Note the narrowed region of the bladder neck (a) above the dilated posterior urethra with valves (b) below, similar to Case I.

Fig. 15. Case IV. The dilated posterior urethra obscures the valves, but the similarity to Cases I and III can be seen.

A suprapubic tube cystostomy was performed, establishing good urinary drainage. This produced some improvement with gradual subsidence of the febrile course and some weight gain. The patient has to date not been considered a suitable candidate for definitive surgery.

Case IV. Baby boy W. was born on January 12, 1959 at Kings County Hospital. On postpartum physical examination a hypogastric mass was noted. Pressure on this mass caused urine to dribble from the urethra. These findings led to a diagnosis of bladder outlet obstruction. The blood level of urea nitrogen was 12 mg. per cent. Urinalysis showed numerous white and red blood cells. A voiding cystourethrogram (Fig. 15) showed a distended bladder and a dilated posterior urethra, suggesting the presence of valves. Reflux into hydronephrotic collecting systems was evident. No definite evidence of function was seen on an excretory urogram. On the fourth day of life a suprapubic tube

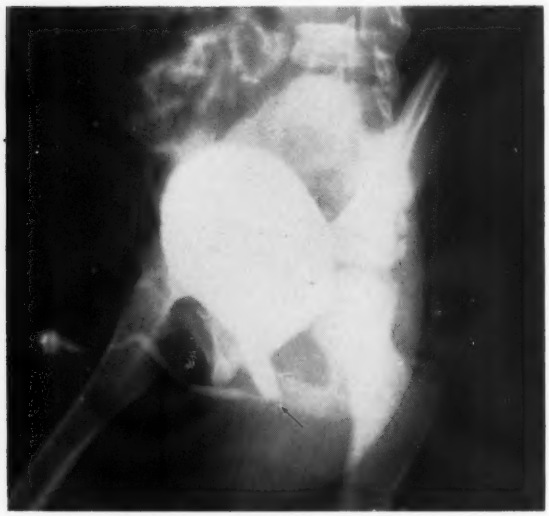

FIG. 16. Neurogenic bladder with funnel-shaped posterior urethra. The arrow indicates the external sphincter.

cystostomy was performed. Postoperatively, the blood urea nitrogen became elevated to 51 per cent. A spiking temperature curve was noted. Urine and blood cultures showed growth of *Streptococcus faecalis*. The sepsis responded to antibiotic therapy and, following correction of electrolyte imbalance, the blood urea nitrogen has returned to normal levels and the child appears to be doing well.

*Comment.* Although this case is not yet proved surgically, we consider the roentgen appearance to be pathognomonic of valvular obstruction.

## DISCUSSION

A review of the literature on congenital urethral valves reveals that the incidence of the disease is decidedly higher in European clinics than in American clinics. Kjellberg, Ericsson and Rudhe[9] reported on 1,155 children with lower urinary tract disease examined at the Karolinska Institute, Stockholm. They found 189 cases with lower tract obstruction of which 52 (27.5 per cent) had congenital valves. In contrast Burns, Pratt and Hendon,[4] reporting 81 cases of lower tract obstruction in children, found 3 (3.7 per cent) with posterior urethral valves. We believe such discrepancies are due to the greater reliance in the European clinics on the voiding cystourethrogram. This roentgenographic technique of examination of the lower urinary tract is not a new one, but it has not achieved sufficient popularity in this country and has been neglected in many radiologic and urologic offices and departments. Instead, reliance is placed on cystoscopic examination which, no doubt, affords much valuable and otherwise unobtainable information. But it is decidedly deficient in certain respects. Like retrograde urethrography the cystoscopic examination requires the retrograde flow of fluid in the urethra. This often causes the delicate valvular folds to float upward and to fold against the wall of the urethra. Thus no obstruction is encountered and the lesion is easily missed. The voiding cystourethrogram duplicates the physiologic conditions of micturition and, in striking contrast, demonstrates the presence of valves. Moreover, the latter is a relatively simple, informative and innocuous method of examination. Neurogenic disease of the bladder is the only condition which in our experience has simulated the appearance of valves on the voiding cystourethrogram. However, the funnel-shaped urethra, dilated to the external sphincter, occurring in this condition (Fig. 16), should not be difficult to differentiate.

Since adding voiding cystourethrography to our studies approximately eighteen months ago, we have found the 4 cases described above. This leads us to the conclusion that we have in the past misdiagnosed a significant number of cases of congenital urethral valves. It is our hope in presenting this material that the wider use of voiding cystourethrography in the examination of the lower urinary tract will produce increased accuracy in the early diagnosis of disease in this region. The roentgenographic appearance of the lower urinary tract should be as familiar as that of the upper urinary tract to the radiolgist and urologist.

## SUMMARY

Voiding cystourethrography should be done on all children with lower urinary

tract symptoms. It can be performed easily even in the newborn infant. Congenital posterior urethral valves will often be missed if cystoscopic examination alone is relied upon for diagnosis. Micturition produces dynamic changes in the appearance of the bladder neck and posterior urethra. Thorough familiarity with these variations is necessary to differentiate them from pathologic changes. Four cases of congenital valves are presented.

Walter A. Griesbach, M.D.
Permanente Medical Group
2425 Geary Boulevard
San Francisco, California

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# ACUTE ATROPHY OF THE THYMUS INDUCED BY ADRENOCORTICOSTEROIDS: OBSERVED ROENTGENOGRAPHICALLY IN LIVING HUMAN INFANTS*

## A PRELIMINARY REPORT

*By* JOHN CAFFEY, M.D., *and* CHARLES DI LIBERTI, M.D.
NEW YORK, NEW YORK

IN 1950, Andersen[1] demonstrated, at necropsy, striking atrophy of lymphoid tissue in the thymus, spleen and lymph nodes of 6 premature infants who had been treated with adrenocorticotrophin (ACTH) to combat retrolental fibroplasia. Silverman and his colleagues[7] reported retardation of growth and increase in the urinary excretion of corticosteroids in the living members of this same group of prematures. Andersen's findings suggested to us that the thymolytic effects of adrenocorticosteroids might be demonstrated in living infants with large thymuses by serial roentgenographic observations of the chest made during and after the administration of corticosteroids. This report records the findings of such a study.

### REPORT OF CASES

CASE I. P. B., a Negro girl, thirteen months of age, had been born prematurely and had always been small. Roentgenograms of the chest disclosed a cardiothymic image widened at all levels but wider on the left than on the right (Fig. 1*A*). Prednisone was given by mouth, 5 mg. 3 times a day for seven days and a total dosage of 105 mg., after which the cardiothymic image was markedly shrunken (Fig. 1*B*). During the treatment, the frontal area of the cardiothymic image had diminished from the original 38 sq. cm. to 29 sq. cm. after treatment (a reduction of 24 per cent), and the cardiothoracic index was reduced from the original 0.63 to 0.52 (a reduction of 17 per cent).

CASE II. A. S., a white boy, four and one-half months of age, had been dyspneic for several days with clinical findings which suggested asthmatic bronchitis to his pediatrician, Dr. E. W. Earl of Pocatello, Idaho. Roentgenograms of the chest disclosed a large thymicocardiac image, widened at all levels (Fig. 2*A*). Prednisone was given in daily doses of 20 mg. for eight days when the thymicocardiac image was substantially reduced (Fig. 2*B*). During

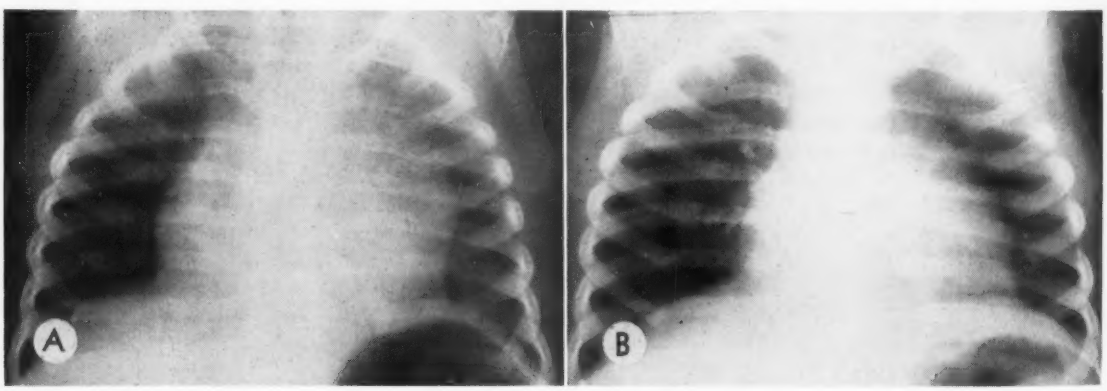


FIG. 1. Case I. Rapid shrinkage of the widened mediastinum following oral administration of corticosteroids to a Negro infant, thirteen months of age. (*A*) Before treatment; (*B*) after 105 mg. of prednisone during seven days.

* From the Departments of Pediatrics and of Radiology, College of Physicians and Surgeons, Columbia University, and the Babies Hospital.
Given in part as the Paul Beaven Honorary Lecture, Rochester, New York, June 2, 1959.

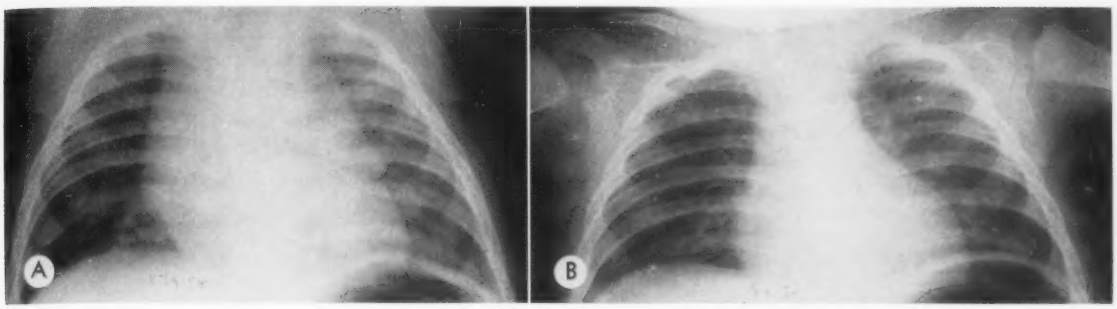

FIG. 2. Case II. Rapid shrinkage of the widened mediastinum after eight days and a total dose of 160 mg. of prednisone given to a white boy, four and one-half months of age. (*A*) Before treatment; (*B*) after treatment (courtesy of Dr. E. W. Earl, Pocatello, Idaho).

treatment, the area of the cardiothymic image had diminished 35 per cent, and the cardiothoracic index was reduced from 0.57 to 0.50 (a reduction of 12 per cent).

CASE III. M. K., a white boy, two months of age, who was having intermittent mild attacks of cyanosis, had roentgenograms of the chest made which demonstrated the right side of the supracardiac image to be widened in a triangular fashion (Fig. 3*A*). Prednisone was given in daily doses of 15 and 20 mg. for twelve days and a total of 200 mg.; the widened supracardiac image was unchanged (Fig. 3*B*). Prednisone was stopped, and in a roentgenogram made seven days later, the right-sided supracardiac widening had disappeared (Fig. 3*C*); but in a roentgenogram made two and one-half months later (Fig. 3*D*), the widened right lobe of the thymus reappeared.

CASE IV. S. S., a white boy, three months of

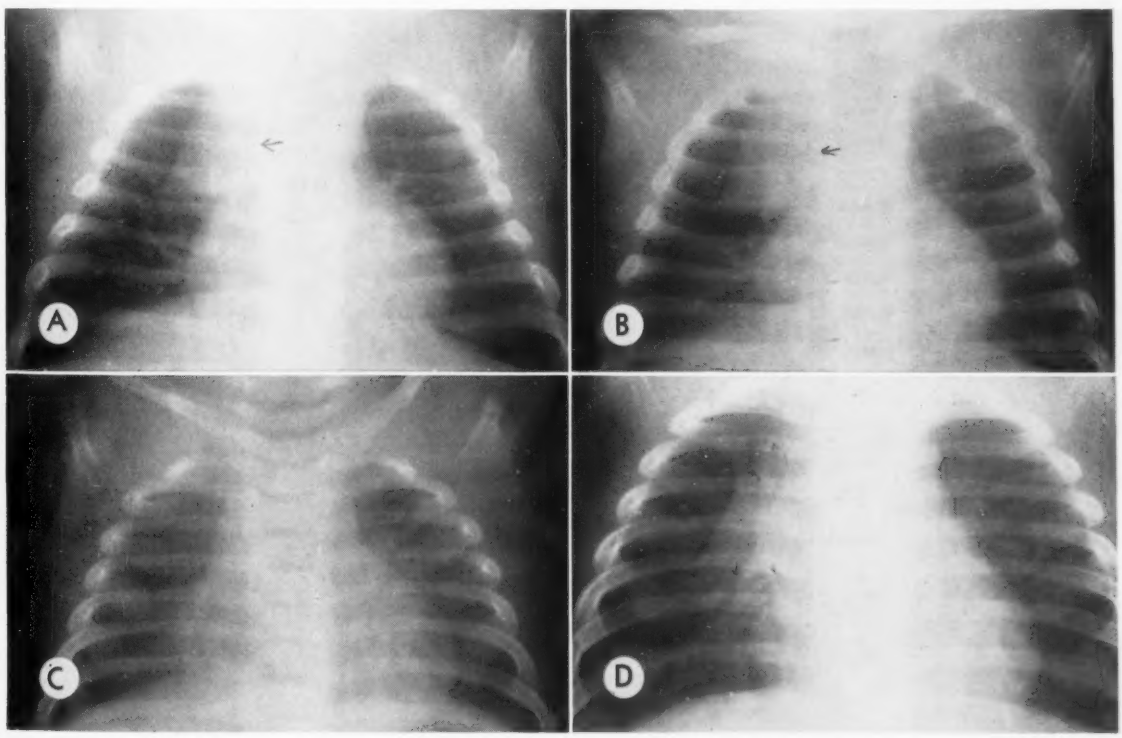

FIG. 3. Case III. Rapid shrinkage of the right side of the widened supracardiac image in a boy two months of age. (*A*) Before treatment; (*B*) persistence of the widening after 200 mg. of prednisone given during twelve days; (*C*) disappearance of widened supracardiac image seven days after *B*, without treatment. (*D*) Regrowth of the right lobe of the thymus two and one-half months after *C*.

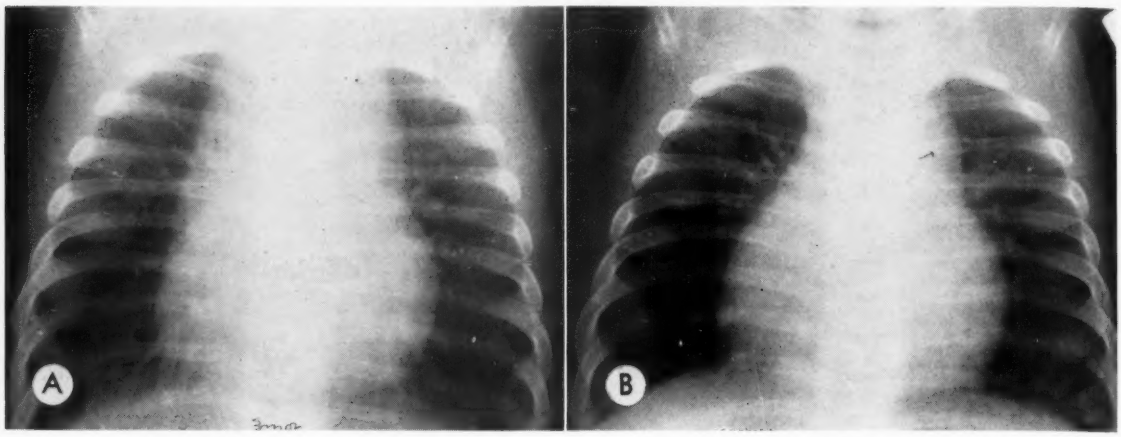


FIG. 4. Case IV. Rapid shrinkage of the widened supracardiac image and visualization of the aorta which had previously been invisible in a boy three months of age, after 172 mg. of prednisone given during twelve days. (*A*) Before treatment; (*B*) after treatment.

age, had had a slowed cardiac beat without cardiac murmur since the fourth week of life. An electrocardiogram showed signs of atrioventricular dissociation without ventricular axis deviation. In the first roentgenogram, made at three months of age, the cardiothymic image was enlarged and widened in a fashion which did not permit satisfactory visualization of the aorta (Fig. 4*A*). The thymicocardiac image contained 40 sq. cm. and the cardiothoracic index equalled 0.56. The major clinical problem was determination of whether the slowing of the heart was due to congenital malformation of the intracardiac septum or to inflammation of the intracardiac septum. Treatment with adrenocorticosteroids seemed indicated to benefit a possible inflammatory lesion in the septum. Prior to treatment, the cardiothoracic index equalled 0.60 and the total area of thymicocardiac image was 46 sq. cm. Prednisone was given for twelve days, in total dosage of 172 mg. In Figure 4*B*, made at the end of this treatment, the cardiothoracic image had shrunken from 46 to 40 sq. cm. (13 per cent) and the cardiothoracic index from 0.60 to 0.56 (6 per cent). Also after the treatment the aorta was visible in its normal position on the left side of the mediastinum which indicated that there was no dextrocardia, but that the true lesion was dextroversion of the ventricles with normally

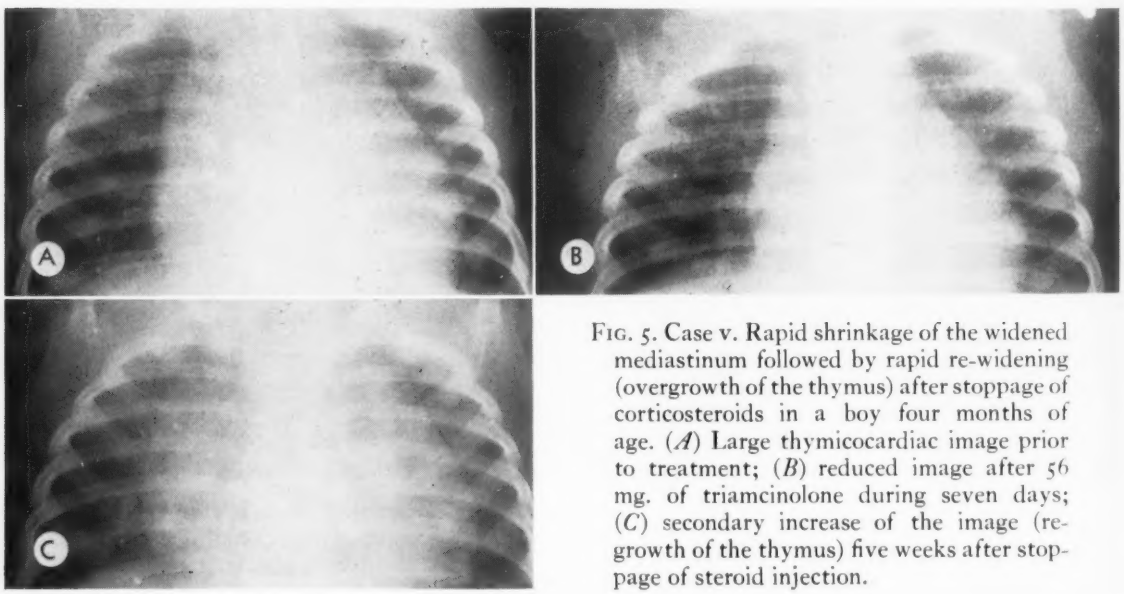


FIG. 5. Case V. Rapid shrinkage of the widened mediastinum followed by rapid re-widening (overgrowth of the thymus) after stoppage of corticosteroids in a boy four months of age. (*A*) Large thymicocardiac image prior to treatment; (*B*) reduced image after 56 mg. of triamcinolone during seven days; (*C*) secondary increase of the image (regrowth of the thymus) five weeks after stoppage of steroid injection.

placed atria—a congenital malformation and not inflammatory disease of the cardiac septum.

Case v. H. T., a white boy, four months of age, was first examined roentgenographically because of laryngeal stridor and the roentgenogram disclosed marked enlargement of the thymicocardiac image (Fig. 5*A*). The cardiothoracic index equalled 0.64 and the thymicocardiac image contained 52 sq. cm. Ten days later triamcinolone was started in oral doses of 4 mg. twice a day for a total of 56 mg. during seven days. At the end of this treatment the thymicocardiac image was strikingly diminished in size (Fig. 5*B*); the cardiothoracic index was reduced to 0.57 (a reduction of 11 per cent) and the total cardiothymic area to 38 sq. cm. (a reduction of 29 per cent). Corticosteroids were stopped, and five weeks later in a third roentgenogram (Fig. 5*C*) the thymicocardiac image was strikingly increased in size and was larger than the image prior to corticosteroid treatment. The cardiothoracic index was now 0.66; the frontal area of the thymicocardiac image now contained 58 sq. cm., a 52 per cent increase over the same image after treatment and a 15 per cent increase over the image prior to starting the steroids.

Case vi. W. B., a girl seventy-one days of age, was examined roentgenographically because of recurrent respiratory infections. The roentgenogram (Fig. 6*A*) disclosed enlargement of the thymicocardiac image; the cardiothoracic index equalled 0.59 and the frontal area of the thymicocardiac image contained 29 sq. cm. Eight days later triamcinolone was started in daily doses of 8 mg. and continued for ten days. A second roentgenogram showed pronounced reduction in the size of the thymicocardiac image (Fig. 6*B*). The cardiothoracic index was diminished to 0.52 (a reduction of 12 per cent) and the area of the cardiothymic image to 21 sq. cm. (a reduction of 28 per cent). Corticosteroids were stopped and fourteen days later (Fig. 6*C*) the area of the cardiothymic image had increased to 32 sq. cm. (an increase of 57 per cent over that demonstrated in Figure 6*B* and 9.6 per cent over the increase shown in Figure 6*A*).

Case vii. J. M., a white boy fifteen months of age, was suspected of having swallowed a thumb tack and roentgenograms were made the same day, which disclosed generalized enlargement of the thymicocardiac image with a localized rounded bulge in the right supracardiac region, which suggested either localized enlargement of the right lobe of the thymus, or of a mediastinal lymph node (Fig. 7*A*). The cardiothoracic index equalled 0.55 and the thymicocardiac image contained 56 sq. cm. Prednisone in doses of 20 mg. daily was given for eight days, a total of 160 mg. A second

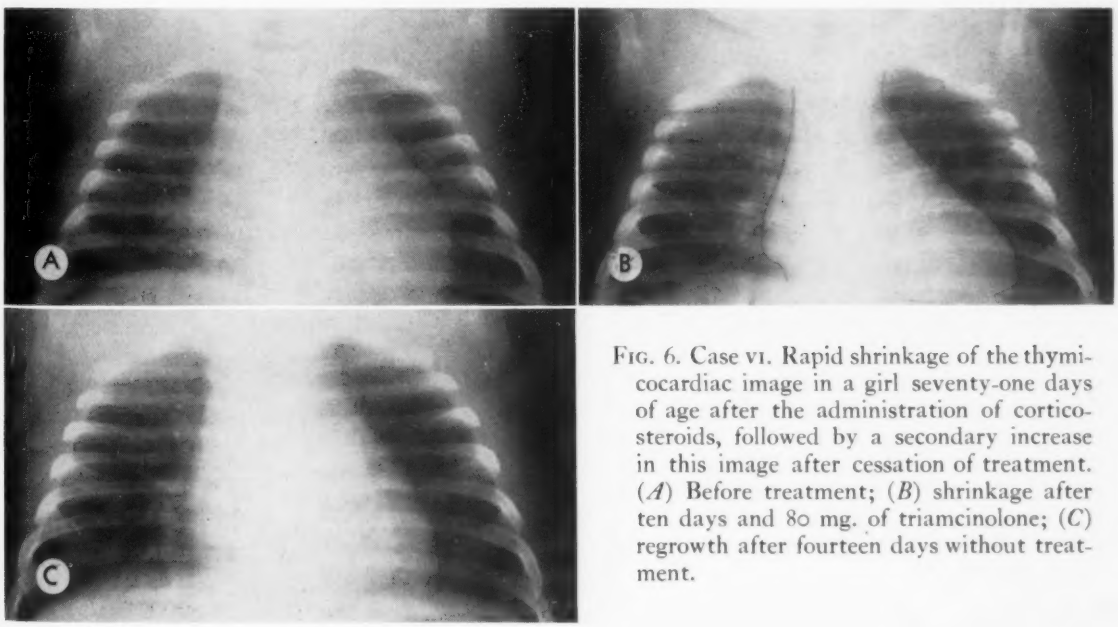


Fig. 6. Case vi. Rapid shrinkage of the thymicocardiac image in a girl seventy-one days of age after the administration of corticosteroids, followed by a secondary increase in this image after cessation of treatment. (*A*) Before treatment; (*B*) shrinkage after ten days and 80 mg. of triamcinolone; (*C*) regrowth after fourteen days without treatment.

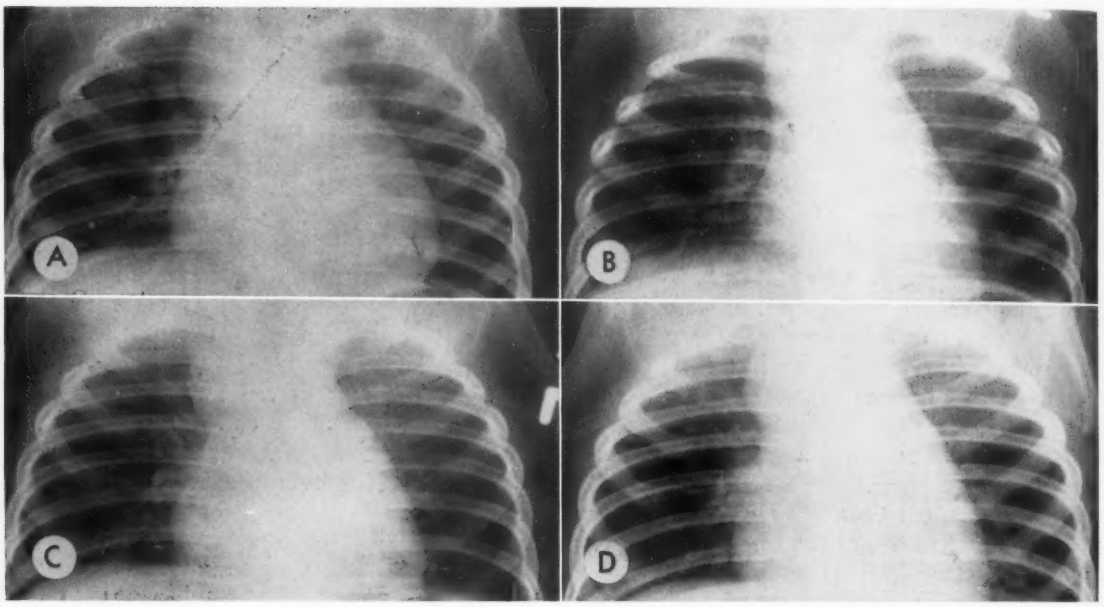


FIG. 7. Case VII. Shrinkage and regrowth of rounded mass (mediastinal lymph node?) on mediastinal border after administration of corticosteroids and then cessation of treatment in an asymptomatic boy fifteen months of age. (*A*) Rounded swelling in right supracardiac image prior to steroids; (*B*) after eight days and 160 mg. of prednisone. The thymicocardiac image has shrunken and the rounded bulge on the right has disappeared; (*C*) nine days after *B* and stoppage of prednisone. The right supracardiac bulge has reappeared and the thymicocardiac image has re-enlarged to its pretreatment size and shape; (*D*) fifteen days after *C* without treatment. The right supracardiac bulge is still large, as is the general thymicocardiac image.

roentgenogram (Fig. 7*B*) made after this treatment disclosed generalized shrinkage of the thymicocardiac image, and the localized bulge on the right side of the mediastinum above the heart had disappeared. The cardiothoracic ratio was reduced to 0.42 (a reduction of 24 per cent) and the area of the thymicocardiac image to 43 sq. cm. (a decrease of 21 per cent in comparison with Figure 7*A*). Prednisone was discontinued and in a roentgenogram made nine days later the right-sided bulge had reappeared and the general cardiothoracic image had increased to 56 sq. cm. (an increase of 30 per cent over that in Figure 7*B* but no increase when compared to Figure 7*A* before treatment). The cardiothoracic index had increased from 0.42 (Fig. 7*B*) to 0.51 sq. cm. (an increase of 21 per cent over that seen in Figure 7*B*). In a roentgenogram (Fig. 7*D*) made nineteen days after Figure 7*C* and twenty-four days after Figure 7*B*, the findings were similar to those in Figure 7, *A* and *C*. In a fifth roentgenogram made six months later and not reproduced here the findings were again practically identical with those in Figure 7*A*.

CASE VIII. B. W., a white boy, was first examined on the second day of life because of repeated "spitting up" and vomiting since shortly after birth. A roentgenogram (Fig. 8*A*) showed the esophagus to be terminated in a blind pouch and the cardiothymic image to be enlarged, with most of the enlargement above the heart. After surgical exploration, the esophageal atresia with tracheo-esophageal fistula was corrected. Seven days later, on the ninth day of life (Fig. 8*B*) and without administration of corticosteroids, roentgenograms of the chest disclosed a sharp shrinkage in the supracardiac and cardiac images. We attributed these changes to natural stress atrophy of the thymus provoked by the stressors, starvation and surgical trauma with anesthesia. The patient then began to thrive and continued to thrive. On the twenty-fourth day of life the thymicocardiac image had increased in size (Fig. 8*C*) and at five months of age the cardiothymic image was strikingly widened at all levels (Fig. 8*D*). This late regrowth and then overgrowth of the thymus, following acute stress atrophy, can probably be best explained by presuming a prolonged

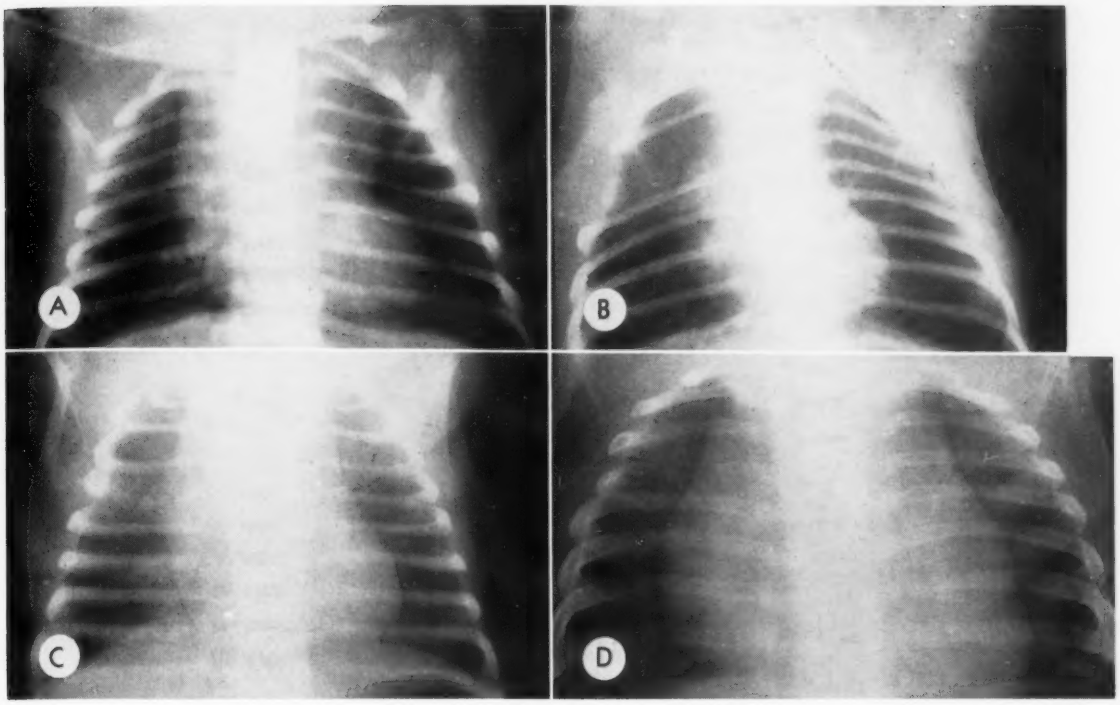


Fig. 8. Case viii. Natural rapid stress atrophy of the thymus followed by regrowth and then overgrowth (*A*) Frontal roentgenogram on the second day of life in which the esophagus is atretic and the supracardiac image widened; (*B*) on the ninth day, seven days after surgical repair of the esophageal atresia and tracheo-esophageal fistula, the supracardiac and cardiac images are greatly reduced in width; (*C*) on the twenty-second day of life, spontaneous increase in both the supracardiac and cardiac images is demonstrated; (*D*) at five months of age, the fused cardiothymic image is greatly widened. The false cardiothoracic index equals 0.64.

exhaustion of the adrenal cortices following the severe stresses encountered during the first days of life which removed the normal inhibition to lymphatic and thymic growth provided by active normal adrenals.

Case ix. M. B., a white boy, was examined on the twenty-third day of life because of cyanotic spells which worsened after feedings. In the first roentgenogram (Fig. 9*A*), the thymicocardiac image was widened on both sides. The cardiothoracic index equalled 0.62, and the total frontal area of thymicocardiac image contained 30 sq. cm. Prednisone was given in 5 mg. doses daily for five days (total dose 25 mg.) and at the end of this treatment the cardiothymic image was not significantly altered (Fig. 9*B*). There were also no changes after twelve additional days of prednisone with a total dose of 145 mg. during seventeen days (Fig. 9*C*), and then after seven more days with a total dose of prednisone of 215 mg. during twenty-four days (Fig. 9*D*). At the end of the steroid therapy the cardiothoracic index equalled 0.60, and the total area of the thymicocardiac image contained 32 sq. cm. The failure of the cardiothymic image to shrink during a long course of corticosteroid in high dosage suggests that the enlargement of the cardiac image was due either to increase in thymic tissue which was insensitive to corticosteroids or to increase in nonthymic tissue, possibly cardiac tissue, which was also insensitive to corticosteroids.

Case x. M. L., a white girl, was first seen at twenty-five months of age with complaints of failure to gain weight and poor appetite. She had never been cyanotic. At physical examination, the heart was enlarged and a hoarse systolic murmur was heard over the precordium. In roentgenograms, the supracardiac image was widened on both sides, and the heart was moderately enlarged with prominent vascular radicles in the lungs. The combined clinical and roentgenographic findings suggested a large interatrial septal defect and anomalous

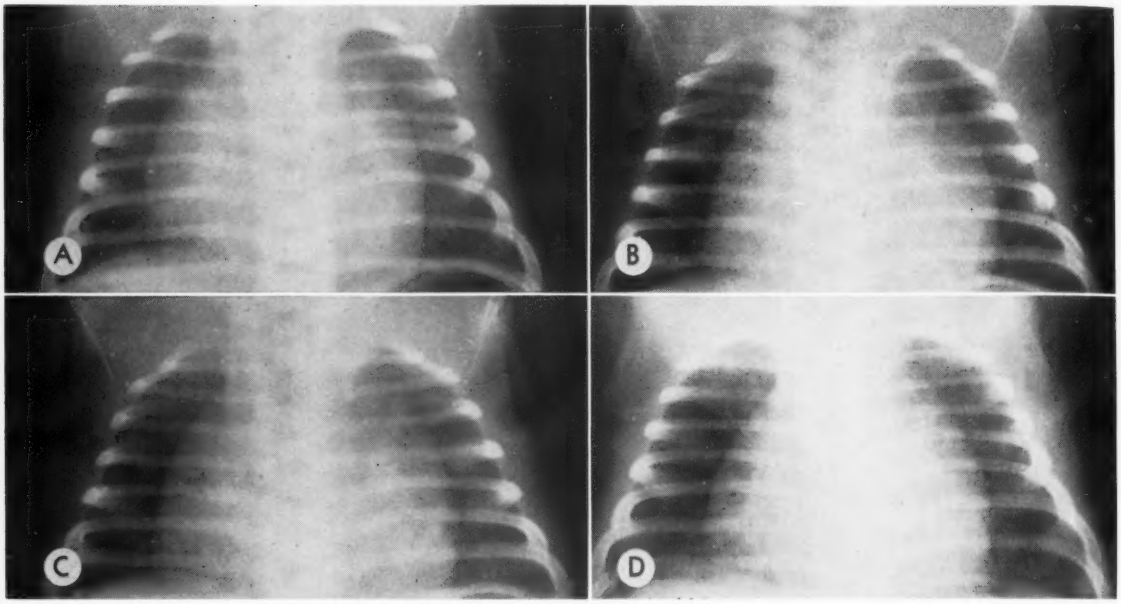


FIG. 9. Case IX. Bilateral widening of the cardiothymic image in a boy twenty-three days of age which did not shrink during twenty-five days of corticosteroid therapy and a total dose of 215 mg. of prednisone. (*A*) Mediastinal widening on the twenty-third day of life before treatment; (*B*) no shrinkage after five days and 25 mg. of prednisone; (*C*) on the fortieth day of life and after seventeen days of prednisone (145 mg.), no changes are noted; (*D*) on the forty-seventh day of life and after twenty-four days of prednisone (215 mg.), the image remains the same. The pretreatment widening of the mediastinum persisted despite large doses of prednisone during three and one-half weeks.

drainage of pulmonary veins. At thirty-two months of age, inconstant cyanosis had developed and the fingers were clubbed. Roentgenograms again showed bilateral widening of the supracardiac image, enlargement of the heart and overcirculation in the lungs (Fig. 10*A*). At thirty-seven months of age, prednisone was started in daily doses of 10 mg. for ten days for a total dosage of 100 mg. Roentgenograms at the end of the treatment (Fig. 10*B*) disclosed bilateral widening of the supracardiac image, enlargement of the heart and overcirculation in the lungs, all similar to the changes present five months before. No untoward clinical effects appeared during or after the treatment. In later opaque venous angiocardiograms the pulmonary veins were ectopic and connected with the left innominate vein, through which most of the venous blood coursed, and a large atrial septal defect was demonstrated

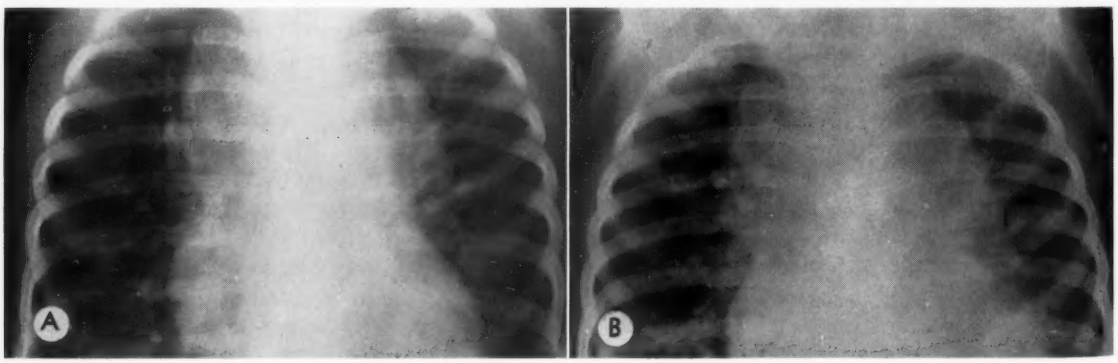


FIG. 10. Case X. Widening of the supracardiac image due to ectopic great veins (proved in opaque venous angiocardiograms) in a girl twenty-five months of age which proved insensitive to adrenocorticosteroids. (*A*) Before treatment; (*B*) after ten days and 100 mg. of prednisone, the supracardiac image is unchanged.

with an enlarged right ventricle and prominent pulmonary arteries. It was manifest that the supracardiac widening, seen in earlier roentgenograms, was due to the ectopic anomalous veins which, of course, were insensitive to the thymolytic effect of corticosteroids.

## DISCUSSION

In 7 of 8 cases (Case I to VII inclusive), in which idiopathic widening of the mediastinum was discovered by chance in random roentgenograms, the oral administration of adrenocorticosteroids caused rapid shrinkage of the mediastinal image. In the eighth case (Case IX) the widened mediastinal image remained unchanged after steroid treatment. We attribute the original widenings of the mediastinal images to enlargement of the thymic lobes in each case, and the shrinkages to the thymolytic effect of adrenocorticosteroids. In 3 cases (Case V, VI and VII), regrowth and overgrowth of the thymus occurred after stoppage of the steroids. In Case VII, a localized swelling on the right side of the mediastinum which we believe probably represented an enlarged, inflamed lymph node also shrank and then regrew following cessation of the steroid. The spleen was not enlarged in any of these patients prior to steroid treatment so that the effect of steroid on splenic size could not be determined by palpation. The findings in Case VIII, we believe, represent natural stress (adrenocorticosteroid) atrophy of the thymus followed by regrowth and overgrowth in a newborn with atresia of the esophagus who suffered from the stresses of starvation, surgery and anesthesia during the neonatal period. It is noteworthy that regrowth and overgrowth in this natural stress atrophy of the thymus is just as marked as after thymic atrophy induced artificially by the administration of cortocosteroids (Fig. 5, *A*, *B* and *C;* 6, *A*, *B* and *C;* and 7, *A*, *B*, *C* and *D*). We have seen similar natural stress atrophy of the thymus in many cases of pneumonia and suppurative pleurisy in infants whose thymuses were originally enlarged. It is probable that stress atrophy of the thymus of this natural type occurs commonly—in all cases probably where the stressors are severe. Rapid regrowth of the thymus also follows shrinkage by exposure to ionizing radiation.[3]

Our infants were all treated while they were out-patients and there were several uncontrollable variables which did not permit accurate determination of the velocity of thymic atrophy, or the optimal or minimal dose required. More adequate studies of these features require repeated frequent roentgenographing of infants at short intervals during several days or weeks, which is potentially hazardous with the roentgen generators currently in use. The dosages of adrenocorticosteroids were largely arbitrary, but we knew they were safe, in view of our treatment of younger infants suffering from infantile cortical hyperostosis with similar doses. In Case I, pronounced atrophy of the thymus was evident after seven days and 105 mg. of prednisone; and, in Case V, after seven days and 56 mg. of triamcinolone. Regrowth after atrophy is also interesting from the standpoint of dose of the steroid and time required for it but it could not be determined in this preliminary study owing to the paucity of repeated roentgen examinations. In Case VI, marked regrowth was already evident fourteen days after stoppage of steroid; and in Case VII, distinct regrowth had occurred only nine days after the steroid was stopped. Under the circumstances of natural shrinkage (Case VIII) regrowth was already evident thirteen days following the stress of surgery and anesthesia, and overgrowth was pronounced five months later.

The degree and completeness of thymic atrophy cannot be evaluated satisfactorily from these roentgenograms because the thymic components of the mediastinal image cannot be differentiated clearly from the cardiac and vascular components, which are insensitive to steroid thymolytic effect. Also, the depth of the cardiothymic image, both before and after skrinkage, cannot be determined because both the ventral and dorsal edges of the thymus are

not clearly defined. The findings do suggest, however, that most of the thymus disappeared during steroid therapy. In cases in which the thymicocardiac image was originally markedly enlarged, frontal areas of the total thymicocardiac image diminished after steroids by 24 per cent, 35 per cent, 29 per cent and 28 per cent even though the larger cardiac component probably did not participate in the shrinkage. The cardiothoracic index was enlarged significantly in all cases with widened mediastinums and then was reduced after corticosteroids by 18 per cent, 14 per cent, 11 per cent, 12 per cent and 23 per cent in different cases. It is manifest that the cardiothoracic index may be an erroneous index of cardiac enlargement during infancy when the thymus tends to be larger, and its reduction during observation may represent stress atrophy of the thymus as well as shrinkage of the heart itself. The use of the cardiothoracic index during the first months of life probably generates more misinformation than fact.

Steroid-induced atrophy of the thymus, observed by us in living human infants, has long been suspected on clinical and morphologic findings in humans, and has been demonstrated experimentally in many small animals. Boyd[2] wrote a comprehensive review of the normal growth of the thymus and its reactions in health and disease in 1932. She states that rapid atrophy of the thymus and lymphoid tissues due to starvation had been reported as early as 1845 by Simon, who called the thymus "a barometer of nutrition and a very delicate one." She (Boyd) also explains the "age involution of Hammar," and his "accidental involution" under such stressors as starvation, fatigue and fever. Santisteban[6] points out that hormonal control of thymolymphatic growth was first suggested by Addison in 1855 when he observed lymphatic hyperplasia in Addison's disease. More complete recognition of the hormonal factors has advanced rapidly since Selye's first account of the "adaptation syndrome" in which he demonstrated thymic atrophy following stimulation of the pituitary-adrenal axis. Dougherty[4] and White proved that the acute thymic and lymphatic atrophy, provoked by starvation, is mediated in large part by excess of adrenocorticosteroids stimulated by adrenocorticotrophin.

Dougherty[4] in a masterful review on the effects of hormones on thymicolymphatic tissues points out that acute lymphatic involution is characterized by actual destruction of the lymphocytes *in situ* in progressive phases of edema, suppression of mitosis, and cellular fragmentation with phagocytosis of the lymphocytic fragments by macrophages. The destructive effect appears to be due to direct action of the corticosteroids on the lymphocytes, because it occurs *in vitro*.

De Asua, according to Santisteban,[6] postulates that thymic and lymphatic tissue hypertrophy occurs during natural states of adrenocortical insufficiency; and when the bone marrow is depressed at the same time, leukemia develops. In this sense, he regards leukemia as a secondary reaction due primarily to the insufficient response of the pituitary axis to stress. The reticuloendothelial system, during states of adrenocortical insufficiency, free of the brake normally imposed by active healthy adrenocorticosteroid apparatus, starts to proliferate, gets out of control and then goes on to wild proliferation and leukemia. In our cases we may have seen the first phase in this cycle due to adrenocorticosteroid inhibition, caused by the oral administration of excess of corticosteroids. The findings in our cases suggest that the thymolytic effect of the corticosteroids, given by mouth, gave out long before the adrenal glands recovered their normal secretory activity because regrowth and overgrowth of the thymus were evident several weeks after stoppage of oral corticosteroids.

Kaplan and co-workers[5] studied the phenomena of regrowth and overgrowth of the thymus in adrenalectomized rats which had also been irradiated. They found that lymphosarcomas often developed in the

overgrown thymus and then appeared widely in other organs and tissues, including the blood-forming organs. Tumor induction however, required total body irradiation, and was strikingly inhibited by shielding the thighs from radiation, or exteriorizing the spleen surgically so that it was outside the field of irradiation. Lymphoma development was practically abolished by thymectomy. They also found that in irradiation-induced leukemia, tumor incidence was increased by adrenalectomy. Cortisone in excess inhibited tumor formation in like circumstances and also in spontaneous leukemia. These authors emphasize that the various types of lymphatic structures differ significantly in their sensitivity to the lytic effects of adrenocorticosteroids—the thymus is the most sensitive, then the lymph nodes, with the spleen the least sensitive. In a discussion of Kaplan and co-workers' paper, Dr. B. L. Baker explained that the lymphoid organs are not identical in their content of lymphoid tissue and therefore do not have the same content of steroid-sentitive cells. The thymus is the most sensitive because it is made up largely of lymphocytes and reticular stroma; the lymph nodes have the same components in lesser degree; the spleen in contrast is much less sensitive because a large portion of the spleen is composed of sinusoids filled with blood which are insensitive to the lytic action of steroids. In the spleen only the malpighian bodies are lymphoid, and therefore steroid sensitive. Also, in discussion of the same paper, Dr. Max A. Goldzieher made the interesting comment: "in observations obtained some time ago in children with so-called enlarged thymuses, intravenous injection of aqueous extract of adrenal cortex produced exactly the same results as irradiation, at least as far as x-ray pictures can show." He did not mention regrowth or overgrowth following stoppage of the adrenocortical water extract, but he did observe the thymicolytic effect of this relatively crude preparation in living human infants, several years before we studied the thymicolytic effects of the more refined corticosteroids, prednisone and triamcinolone.

## SUMMARY AND CONCLUSIONS

Oral administration of adrenocorticosteroids to human infants with enlarged thymuses caused rapid thymic atrophy. Rapid thymic regrowth and overgrowth occurred following stoppage of the steroid in 3 cases in which adequate follow-up observations were made. In one case of natural stress thymic atrophy, regrowth and overgrowth occurred in the same fashion as in artificially induced thymic atrophy.

When the thymic lobes are large, they usually overlap and their roentgenographic images fuse with the lateral edges of the heart on one or both sides. Under such circumstances the transverse diameter of the heart cannot be measured accurately and when such measurement is attempted, it leads to a spurious large value. This in turn leads to an erroneously enlarged cardiothoracic index. For these reasons, both the absolute values for transverse diameter of the heart and the cardiothoracic index are often misleading and lead to mistaken diagnosis of cardiomegaly. True values for heart size can be obtained after shrinkage of the large thymus with corticosteroids. The true shape of the heart is also masked by enlarged thymic lobes in case of the normal heart and the deformed heart. Thymic shrinkage by steroids enables one to visualize the real cardiac contours, and vascular images above the heart.

Corticosteroid shrinkage of triangular widening of the mediastinum makes possible the differentiation of a conspicuous triangular right lobe of the thymus from atelectasis of the right upper lobe of the lung, segmental pneumonic consolidation of the right upper lobe, and loculated mediastinal pleurisy.

Fluctuations in the size of the mediastinum observed during and after the oral administration of corticosteroids explain, in part at least, the causal mechanism for the great variations in the size of the

mediastinum found commonly in roentgenograms of healthy and sick infants. It seems likely that natural stress atrophy stimulates the pituitary-adrenal axis to the excess secretion of steroids, which produce acute thymic atrophy at first, followed by regrowth and overgrowth. The great variation in the size of the thymus in newborns suggests that many of them have suffered natural steroid stress atrophy of the thymus prior to birth, and some regrowth and overgrowth following the stress atrophy. In this often puzzling problem, we should keep in mind the possibility of hidden internal stresses which are difficult and sometimes impossible to determine from the history.

The marked and possibly harmful destruction of the thymus and all lymphatic tissues by adrenocorticosteroids should be considered in all of the prolonged regimens of corticosteroids used in the treatment of such diseases as rheumatoid arthritis, rheumatic fever, nephrosis, the chronic allergic states and mesenchymal diseases such as lupus erythematosus. It is possible that the overgrowth of thymus, which often follows acute thymic atrophy induced by corticosteroids, may in special circumstances lead on to malignant lymphoid tumors and leukemia.

John Caffey, M.D.
Babies Hospital
167th Street and Broadway
New York 32, New York

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# ROENTGEN THERAPY OF CERTAIN COMPLICATIONS OF ACUTE LEUKEMIA IN CHILDHOOD*

*By* GIULIO J. D'ANGIO, M.D., AUDREY E. EVANS, L.R.C.P. and S., E., *and* ANNA MITUS, L.R.C.P., M.R.C.S.
BOSTON, MASSACHUSETTS

THE course of acute leukemia in childhood is punctuated by many complications.[3,8,19,20,24,26] The control of these secondary manifestations assumes greater importance because of the prolongation of life possible since the introduction of antileukemic chemotherapy.[6] While the basic management of acute leukemia is chemotherapeutic, roentgen therapy can play a significant role in helping to preserve the comfort and well-being of these patients.[10,13,14,18,23,25]

Between 1953 and 1957, 372 children under fourteen years of age with acute leukemia, exclusive of those with an initial lymphosarcomatous phase, were treated in The Children's Cancer Research Foundation of The Children's Medical Center. One hundred and ninety-nine were boys and 173 were girls. Of these patients, 95 (55 boys and 40 girls) received roentgen therapy for some secondary manifestations of their illness (Table I). Many of the remaining children had similar but less severe complications and did not require roentgen therapy. General conclusions regarding the incidence of various complications of acute leukemia cannot be inferred from the figures to be presented since they are based on a selected group of patients. Of the complications amenable to roentgen treatment, those involving the central nervous system, the bones and joints, the urinary tract, and the skin and mucous membranes were the most common. Although many of these manifestations were obvious clinically, lumbar puncture, roentgenographic examination, biopsy and other special studies were employed when required for confirmation. All of the children were receiving chemotherapy throughout the course of their disease. There did not appear to be any significant relation between the agent in use and the development of any of the complications reviewed.

In assessing the results of treatment, the total number of episodes of each of the various complications occurring in the children irradiated was considered. Thus, if a child developed three different manifestations, or the same one on a similar number of occasions, three episodes were listed as having taken place. The response to therapy was divided into four categories: excellent, good, fair, and poor.

The technical factors most frequently employed for roentgen therapy during the period covered by this study are shown in Table II. Techniques in use at present are those designated B, E, and F. The others represent those factors available on older apparatus, since replaced. Doses are given in terms of depth dose, expressed in roentgens. Conversion from roentgens to rads can be accomplished by the use of Table III, abstracted from the National Bureau of Standards Handbook No. 62.[17] The exposure dose (in roentgens) is multiplied by an appropriate factor to obtain the average absorbed dose (in rads). Depth doses administered were calculated for the midplane of the area treated, except for very superficial lesions where the depth dose is practically equivalent to the skin dose.

## CENTRAL NERVOUS SYSTEM COMPLICATIONS

The use of roentgen therapy in the management of these complications has already been described[19,25,29] and the pathology presented in much detail.[4,9,11,27,29] Briefly, the

* From the Department of Radiology, the Department of Medicine, and The Children's Cancer Research Foundation, The Children's Medical Center, and the Department of Radiology, Harvard Medical School, Boston, Massachusetts. Presented at the Fifty-ninth Annual Meeting of the American Roentgen Ray Society, Washington, D. C., September 30–October 3, 1958.
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TABLE I

COMPLICATIONS OF ACUTE LEUKEMIA TREATED BY ROENTGEN THERAPY (SELECTED GROUP OF 95 PATIENTS IN 372 CONSECUTIVE CHILDREN WITH ACUTE LEUKEMIA)

| Site of Involvement | No. of Patients | Per Cent of Total Group | Sex | | Age at Onset of Disease (yr.) | | Interval between Onset of Disease and First Complication (yr.) | | Interval between Recurrences (yr.) | | Total No. of Episodes of Each Complication | Hematologic Status at Time of Each Episode | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | M | F | Average | Range | Average | Range | Average | Range | | Remission | Relapse† |
| Central Nervous System | 37 | 9.9 | 20 | 17 | 5-3/12 | 7/12 to 12-6/12 | 9/12 | P.S.* (6 pts) to 3-3/12 | 3/12 | 1/52 to 1-3/12 | 86 | 46 | 40 |
| Bone and Joint | 25 | 6.7 | 13 | 12 | 5-1/12 | 2 to 12-6/12 | 11/12 | 1/12 to 3-1/12 | 1/12 | 1/52 to 4/12 | 83 | 21 | 62 |
| Skin and Mucous Membranes | 19 | 5.1 | 13 | 6 | 6 | 1-6/12 to 13-6/12 | 1-1/12 | P.S.* (1 pt) to 3-5/12 | no recurrences | | 26 | 1 | 25 |
| Urinary Tract | 18 | 4.8 | 13 | 5 | 4-6/12 | 5/12 to 13-6/12 | 1-5/12 | P.S.* (1 pt) to 8-2/12 | 1/52 (one recurrence only) | | 19 | 0 | 19 |
| Other | 11 | 3.0 | 5 | 6 | 5-10/12 | 1-3/12 to 13-6/12 | 1-7/12 | P.S.* (1 pt) to 8-2/12 | no recurrences | | 14 | 0 | 14 |

* P.S = Presenting symptom.
† Including those prior to first remission.

TABLE II

| Technique | kv. | ma. | Added Filter | Half Value Layer | Target Skin Distance (cm.) | r/min. (as measured in air) |
|---|---|---|---|---|---|---|
| A | 125 | 5 | 1 mm. Al | 3.1 mm. Al | 18 | 160 |
| B | 100 | 25 | 1 mm. Al | 3.8 mm. Al | 50 | 49 |
| C | 200 | 10 | $\frac{1}{2}$ mm. Cu+1 mm. Al | 1.3 mm. Cu | 50 | 23 |
| D | 200 | 10 | 1 mm. Cu+1 mm. Al | 1.6 mm. Cu | 50 | 17 |
| E | 250 | 15 | $\frac{1}{2}$ mm. Cu+1 mm. Al | 1.7 mm. Cu | 50 | 92 |
| F | 250 | 15 | 0.4 mm. Sn+$\frac{1}{4}$ mm. Cu+1 mm. Al | 2.5 mm. Cu | 50 | 50 |

Table III

AVERAGE ABSORBED DOSE PER ROENTGEN OF EXPOSURE IN DIFFERING MATERIALS FOR VARIOUS QUALITIES OF ROENTGEN RADIATION

| Half Value Layer | f = (rad/r) | | |
|---|---|---|---|
| | Water | Muscle | Bone |
| 5.5 mm. Al | 0.91* | 0.94* | 3.10* |
| 1.0 mm. Cu | 0.95 | 0.95 | 1.76 |
| 2.0 mm. Cu | 0.96 | 0.96 | 1.42 |
| 1.7 mm. Cu | 0.96 | 0.96 | 1.44 |
| 2.5 mm. Cu | 0.97 | 0.96 | 1.22 |

* No major inaccuracy will result in the values calculated for absorbed dose (in rads) if these factors are applied to exposure doses (in roentgens) obtained with half value layers of 3 or 4 mm. of aluminum.

findings at autopsy are those of perivascular cuffing, with leukemic infiltration of the dura, the meninges, the brain substance and, on occasion, the cranial nerve nuclei and roots (Fig. 1). The spinal cord and the dorsal and ventral nerve roots may be similarly involved.[29] Commonly, there are subdural, subarachnoid and cerebral hemorrhages, ranging from petechial to massive.

Thirty-seven children, about half of them in hematologic remission,[7] had neurologic manifestations at some time during the course of their illness (Table 1). Ten of these patients developed complications as a presenting symptom or before specific chemotherapy was started. The majority developed headache and vomiting suggestive of increased intracranial pressure or meningeal irritation. Most of them had papilledema and widened sutures on roentgenographic examination of the skull. Usually, the symptoms and clinical signs correlated, although sometimes increased intracranial pressure occurred without relevant complaints. All of the children except 5 had lumbar punctures at the time the complications first developed. Two to 5 cc. of cerebrospinal fluid was removed. The pressure varied from normal levels to more than 600 mm. of water, the cells from 0 to 10,000 mononuclear cells/cu. mm., the protein from 10 to 584 mg./100 cc., and the sugar from 8 to 112 mg./100 cc. Routine bacterial cultures showed no growth. The findings on lumbar puncture did not differ significantly between patients in remission and those in relapse. The cerebrospinal fluid usually returned to normal when the symptoms subsided, but, in a few children who were symptom-free, a steady increase in protein content and blood cell count was noted. The cells present in the spinal fluid were mononuclear in type and could be identified as young forms or blasts. The concept of a "blood-brain barrier" is an attractive explanation for the persistence of these abnormal forms in the spinal fluid of a child in hematologic remission.[25,28] Other mechanisms, however, must also be operative since, in an occasional child, apparently in hematologic remission, leukemic aggregates have been found in other tissues, such as the bowel wall or skin.

The complications in the 37 children with central nervous system involvement are listed below.

| Central Nervous System Complications | No. of Patients |
|---|---|
| Increased intracranial pressure | 17 |
| Increased intracranial pressure with cranial nerve palsies | 7 |
| Increased intracranial pressure with spinal cord lesions | 3 |
| Increased intracranial pressure with proptosis | 2 |
| Increased intracranial pressure with tremor of the limbs | 2 |
| Isolated cranial nerve palsies | 2 |
| Personality change with mental depression | 1 |
| Isolated spinal cord lesion | 1 |
| Excessive appetite and weight gain | 1 |
| Proximal polyneuropathy | 1 |
| Total | 37 |

The child with excessive appetite and weight gain, similar to the patients described by Sansone[21] and Sullivan,[25] did not improve after two separate courses of roentgen therapy directed to the brain.

The patient who had signs of a proximal polyneuropathy developed generalized muscular weakness at the onset of her disease and also showed signs of increased

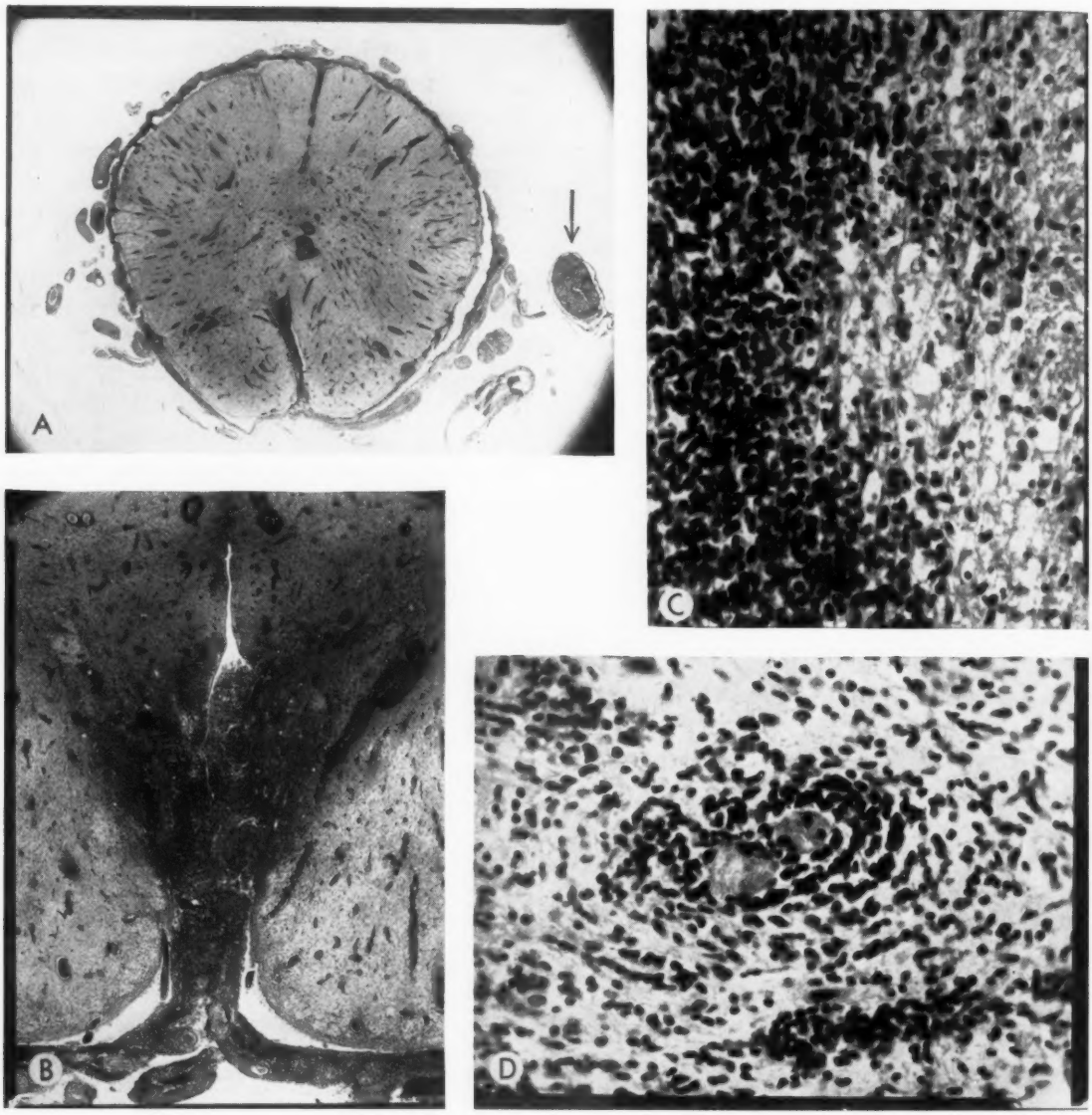


FIG. 1. (*A*) Cross section of cervical cord (×15) of a ten year old girl with acute leukemia. There is marked leukemic infiltration of the subarachnoid space and the perivascular regions with extension into the cord. The arrow points to a nerve root showing perineurial involvement. (*B*) Medulla (×50). Leukemic cells are visible in the subarachnoid space, extending directly into the medullary substance and the fourth ventricle. Perivascular cuffing is also present. (*C*) Medulla (×525). Appearance of the margin of the dense cellular infiltrate seen in *B*. There is invasion of normal brain substance by leukemic cells. (*D*) Medulla (×575). Detailed view of the perivascular cuffing.

intracranial pressure. These unusual findings were thought to be the result of meningeal infiltration and involvement of the nerve roots in the cervical region. She was treated with roentgen radiation, receiving 405 r (depth dose) to the posterior fossa and 600 r (depth dose) to the cervical spine, with complete clearing of her symptoms. At autopsy, extensive leukemic infiltration of the brain, spinal cord, and of their coverings was found.

In general, central nervous system complications were managed by an initial lumbar puncture. Because focal intracranial leukemic masses are rare and the increased pressure is evenly distributed, the danger

TABLE IV

CENTRAL NERVOUS SYSTEM COMPLICATIONS
RESPONSE TO TREATMENT

| | Total | Excellent | | Good | | Fair | | Poor | | Not Evaluated |
|---|---|---|---|---|---|---|---|---|---|---|
| | | No. of Episodes | Per Cent | No. of Episodes | Per Cent | No. of Episodes | Per Cent | No. of Episodes | Per Cent | No. of Episodes |
| I. Lumbar Puncture | 62* | 3 | 7.0 | 20 | 46.5 | 3 | 7.0 | 17 | 39.5 | 19 |
| II. Roentgen Therapy | 24 | 5 | 20.8 | 12 | 50.0 | | | 7 | 29.2 | |
| III. Combined Lumbar Puncture and Roentgen Therapy | 38 | 14 | 38.9 | 13 | 36.1 | 3 | 8.3 | 6 | 16.7 | 2 |

* Of these 62 episodes, roentgen therapy was added in 38 within a few days. The results of this combination were again evaluated and are given in line III.

of herniation of the cerebellar tonsils is small. Nonetheless, lumbar puncture was performed with care and only a few milliliters of fluid was allowed to escape slowly. If there was little improvement or early recurrence after lumbar puncture, the child received roentgen therapy to the skull. At times, roentgen treatment was given without prior lumbar puncture. The response to these different methods of treatment is given in Table IV.

It was of considerable interest to find that the condition of approximately one-half of these patients improved after lumbar puncture alone. The mechanism by which relief is obtained is not known. Possibly, the removal of fluid, together with any extradural leakage which may occur, is sufficient to relieve the pressure and restore the normal dynamics. In view of these observations, it would seem that the results of antifolic compounds administered intrathecally after lumbar puncture[21,28] must be evaluated with caution. In the 4 children of our group treated by this method, the results were no better than after lumbar puncture alone. Roentgen therapy of the skull, with or without prior lumbar puncture, produced more lasting benefit in a higher percentage (70–75 per cent) of these children. The best results were obtained with treatment early in the development of the complication and before irreversible changes, such as permanent cranial nerve palsies, occurred. In our experience, the method of choice is lumbar puncture, followed by roentgen therapy if the symptoms persist or recur within a few days.

Return of symptoms was noted frequently, the 37 children having a total of 86 episodes. Twenty-two had at least one recurrence; of these, 14 had 2, and 6 had 3 or more. The average interval of relief before recurrence following lumbar puncture alone was one month, except for the unusually long response of fifteen months in one patient. Following roentgen therapy alone, the average period before recurrence was three months, and following combined lumbar puncture and roentgen therapy, it was four months. Should symptoms recur, re-treatment in small dosage can be given with hope of success. Radioresistance does not seem to develop in these patients. For example, one child responded well to each of six separate courses of roentgen treatment over a period of one year.

In a few patients, an increase in discomfort and vomiting developed after lumbar puncture and roentgen therapy, and convulsions occurred occasionally. The time interval between treatment and the onset of symptoms ranged from a matter of hours to two days. It was difficult to be certain

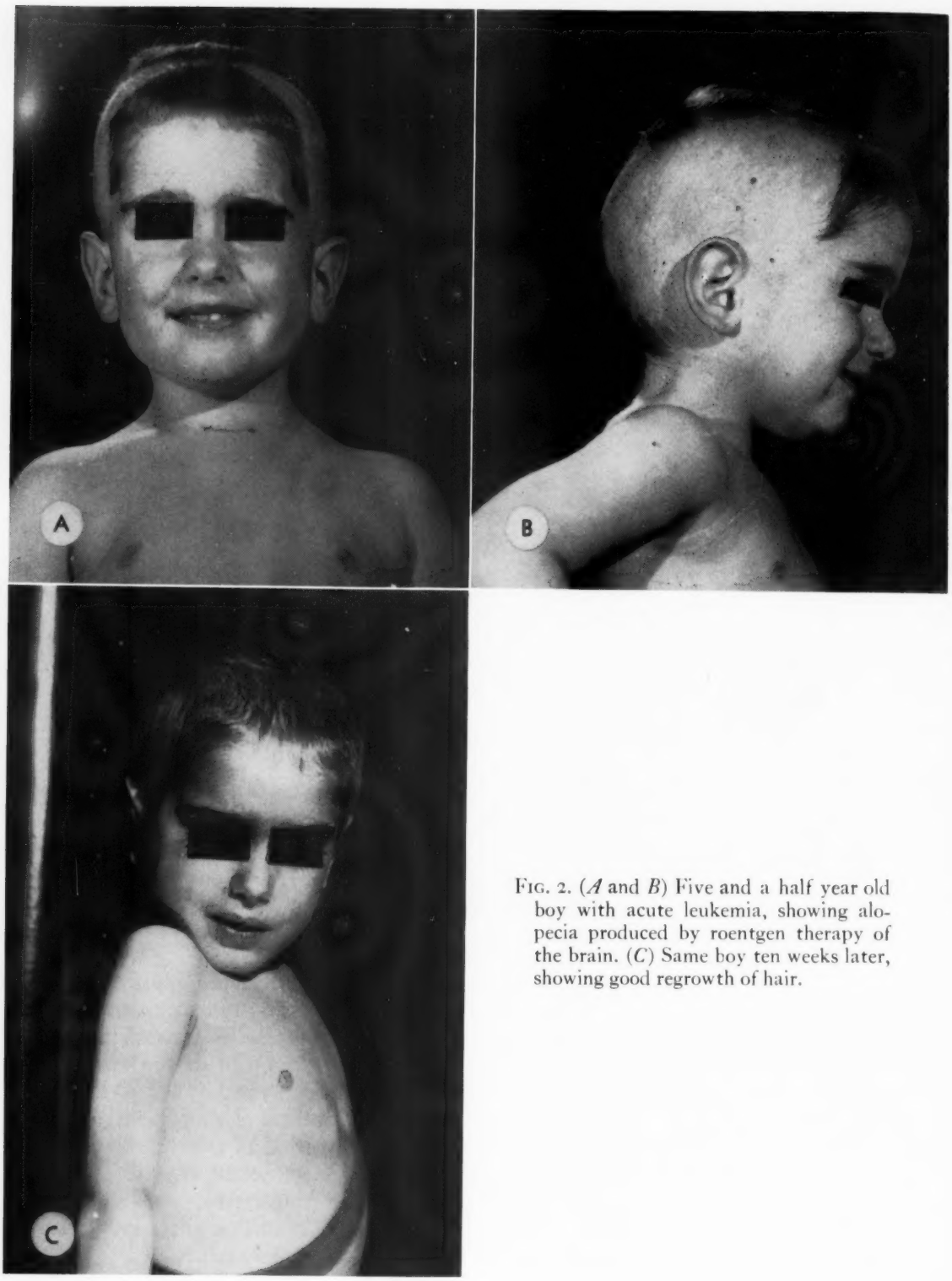


FIG. 2. (*A* and *B*) Five and a half year old boy with acute leukemia, showing alopecia produced by roentgen therapy of the brain. (*C*) Same boy ten weeks later, showing good regrowth of hair.

in the individual patient if the change had been the result of the lumbar puncture, initial edema consequent on roentgen therapy, or extension of the disease process. It is our opinion that progression of symptoms in the face of a cautious first dose of roentgen therapy indicates further leukemic involvement. It has been our practice, therefore, to continue treatment. Epilation (Fig. 2) was noted in many of the patients receiving skull irradiation. Since the roentgen ray dose to the skin was small, loss of hair was not expected to occur so consistently on this basis. It is believed, therefore, to be a response to the combination of irradiation and chemotherapy. In most patients the hair grows back within a few months.

Four children, 3 of them at the onset of their disease, had muscular weakness or paralysis, considered to be the result of spinal cord compression or nerve root involvement by the leukemic process.[29] Another had sensory signs on a similar basis. Laminectomy was considered in 2 of these patients, but was not required. Cautious roentgen therapy was administered to all 4 children with the satisfactory results indicated in Table v.

Roentgen therapy currently is administered using techniques E or F (Table II). When the brain or its coverings are thought to be involved, two opposing lateral skull portals sufficient in size to include most of the brain are ordinarily employed. The eye is shielded directly with 2 mm. of lead placed over the eyelids, and the ear lobe is protected in similar fashion, or folded down and held out of the roentgen-ray beam with cellophane tape. These precautions are taken since repeated courses of roentgen therapy have been required in the same individual, and radiation reactions in these areas are thus avoided. When the symptoms are referable to the spinal cord, the segments involved, with 2 to 3 cm. margins above and below, are treated through a single posterior portal, or 2 posterior oblique fields. An initial dose of 25 to 200 r (air) is given. The exposure varies inversely with the likelihood of causing damage through local edema induced by irradiation. For example, a dose of 25 r (air) is given if the papilledema is marked, the sutures widely split as evidenced roentgenographically, and nausea and vomiting are pronounced. If, however, the manifestation is an isolated cranial nerve palsy without other signs, 200 r in air is administered on the first day. A total depth dose of approximately 450 r is given, although treatment is stopped earlier should a satisfactory response be obtained. Higher doses are now rarely administered. As can be seen from Table v, the response remained

TABLE V

CENTRAL NERVOUS SYSTEM COMPLICATIONS
RESULTS OF ROENTGEN THERAPY

| Site of Involvement | Course of Treatment | Excellent | | | | Good | | | | Fair | | | | Poor | | | | Total |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | Episodes | | Depth Dose | | Episodes | | Depth Dose | | Episodes | | Depth Dose | | Episodes | | Depth Dose | | |
| | | No. | Per Cent | Average (r) | Range (r) | No. | Per Cent | Average (r) | Range (r) | No. | Per Cent | Average (r) | Range (r) | No. | Per Cent | Average (r) | Range (r) | |
| Skull | First | 10 | 45.5 | 481 | 140 to 964 | 7 | 31.8 | 574 | 192 to 1,390 | 2 | 9.1 | 273 | 186 and 360 | 3 | 13.6 | 892 | 340 to 1,190 | 22 |
| | Second | 5 | 27.8 | 560 | 342 to 1,100 | 8 | 44.4 | 590 | 120 to 1,150 | 1 | 5.6 | 868 | | 4 | 22.2 | 268 | 70 to 570 | 18 |
| | Third | 5 | 33.3 | 326 | 142 to 394 | 6 | 40.0 | 368 | 136 to 659 | | | | | 4 | 26.7 | 353 | 260 to 432 | 15 |
| | Total | 20 | | | | 21 | | | | 3 | | | | 11 | | | | 55 |
| Spine | First | | | | | 4 | 57.1 | 359 | 196 to 600 | | | | | 3 | 42.9 | 537 | 350 to 919 | 7 |

TABLE VI

BONE AND JOINT PAIN
RESULTS OF ROENTGEN THERAPY

| | Excellent | | Good | | Fair | | Poor | | Total |
|---|---|---|---|---|---|---|---|---|---|
| | First Episode of Pain | Recurrent Pain | First Episode of Pain | Recurrent Pain | First Episode of Pain | Recurrent Pain | First Episode of Pain | Recurrent Pain | |
| No. of Sites Treated | 12 (14.6%) | 1 (1.2%) | 50 (60.2%) | 8 (9.6%) | 8 (9.6%) | 2 (2.4%) | 1 (1.2%) | 1 (1.2%) | 83 |
| Depth Dose Average (r) | 405 | 270 | 438 | 400 | 383 | 487 | 352 | 570 | |
| Range (r) | 75–833 | | 53–1,438 | 259–848 | 137–580 | 455 and 618 | | | |

poor in some patients, even though roentgen therapy beyond this level was attempted at times.

### BONE AND JOINT PAIN

Metaphyseal separations and fractures, including compression of the vertebral bodies,[1,5] are common in children with acute leukemia and require appropriate orthopedic care. Less specific aches and pains attributable to underlying infiltration of the bony skeleton and soft tissues also occur often, sometimes as a presenting symptom,[19,20,22,26] and may develop regardless of the hematologic status of the patient. Appropriate analgesics usually are adequate for control of discomfort but sometimes the symptoms are so marked and persistent as to prove incapacitating. For example, the child may refuse to walk and becomes bedridden even though under satisfactory hematologic control and in good general condition. The roentgenograms usually show the bone and soft tissue changes associated with acute leukemia in children, but may fail to demonstrate more pronounced findings in the symptomatic area. Despite the absence of obvious cause for local discomfort, it is our practice to proceed with roentgen therapy.

Twenty-five children received roentgen treatment for this complication (Table I). Several developed successive symptoms in more than one area. The sites affected were: knee, 21; spine, 12; ankle, 11; hip, 8; foot, 6; elbow, 3; shoulder, 2; wrist, 2, and various others, 6. A second episode of pain occurring in the same site was noted 9 times, while there were 3 instances of a further recurrence of symptoms. Roentgen therapy was administered, using techniques C, D, E and F (Table II). Daily doses ranged from 50 r (air) to 300 r (air) and were given at intervals ranging from twenty-four hours to one week. Satisfactory control of symptoms was obtained in 85 per cent of episodes treated (Table VI) after relatively small amounts of irradiation, the average dose being less than 450 r (depth). Pursuing therapy to higher dose levels did not appear to improve the results.

It is our current practice to use techniques E and F (Table II). The portals are generous in size and designed to include both the bones and surrounding soft tissues of the involved area. An initial air dose of 200 r to 300 r is given, depending on the thickness of the part, and results of the first treatment are assessed after forty-eight hours. Not infrequently, a single dose will provide complete control of symptoms in twenty-four hours or less, in which case treatment is not repeated. If a depth dose of 450 r fails to result in improvement, therapy is usually terminated.

### SKIN AND MUCOUS MEMBRANE COMPLICATIONS

Infiltration of the skin[20,26] and mucous membranes[2,15] by leukemic cells is associated

with a variety of local lesions ranging from nodular masses in the superficial layers, to hemorrhagic, necrotic, infected ulcers which can extend to involve deeper structures. Cutaneous lesions commonly affect the fingers and toes. Small pustules develop along the surfaces of the digit involved, particularly over the nail beds. These then break down and become infected. Hot soaks, antibiotics, and incision and drainage may not be effective, healing apparently being delayed by the local leukemic process (Fig. 3). Gingival infiltrates and those in the mucosa of the mouth follow a comparable course, although the initial lesion of the buccal mucosa commonly takes the form of a small nodule. The children bite on these and ulceration and infection follow swiftly, with large, foul, deep craters resulting frequently. Roentgen therapy has been found to be of considerable assistance in the control of these local lesions. Nineteen patients with a total of 26 lesions were thus treated for this complication (Table I). In approximately one-half of the children, a gratifying response to roentgen therapy was obtained, frequently within a few days (Table VII). Each of the techniques listed in Table II was used for the treatment of these lesions, their depth determining the choice of half value layer. Doses of 200 r to 300 r (air) were given daily, or every other day, the total amount depending on the response. In some in-

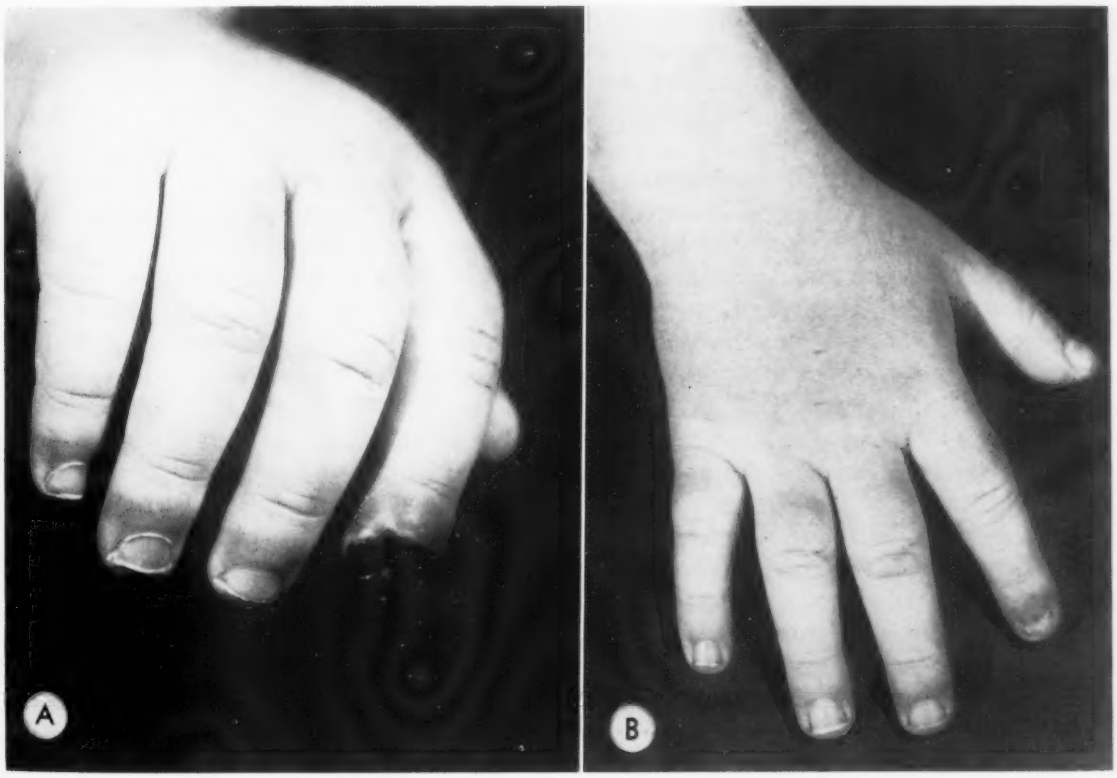


FIG. 3. (*A*) Right index finger of a four year old boy with acute leukemia, showing the result of leukemic infiltration and secondary infection. Despite hot soaks, antibiotics, and surgical procedures including incision and drainage, the process continued to advance. Early gangrene was evident at the time of the photograph. The finger then received roentgen treatment. A total skin dose of 448 r (half value layer 3.1 mm. Al) was given in two treatments forty-eight hours apart. (*B*) Right index finger, appearance three and a half months later. The finger responded promptly to irradiation. After one week, the infection had subsided and satisfactory healing was clearly established. The nail bed had been damaged by the infection so that there was a delay in regrowth of the nail.

TABLE VII

SKIN AND MUCOUS MEMBRANE COMPLICATIONS
RESULTS OF ROENTGEN THERAPY

| Site of Involvement | Excellent | | | Good | | | Fair | | | Poor | | | Not Evaluated | | Total |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | No. of Lesions | Depth Dose Average (r) | Depth Dose Range (r) | No. of Lesions | Depth Dose Average (r) | Depth Dose Range (r) | No. of Lesions | Depth Dose Average (r) | Depth Dose Range (r) | No. of Lesions | Depth Dose Average (r) | Depth Dose Range (r) | No. of Lesions | Depth Dose (r) | |
| Skin | 3 (23.1%) | 416 | 256 to 600 | 5 (38.4%) | 459 | 180 to 565 | 2 (15.4%) | 151 | 118 and 183 | 3 (23.1%) | 358 | 311 to 420 | | | 13 |
| Mucous Membranes of Mouth | | | | 5 (50.0%) | 247 | 183 to 355 | 1 (10.0%) | 190 | — | 4 (40.0%) | 428 | 190 to 752 | 1 | 53 | 11 |
| Anus and Perianal Tissues | | | | 1 (50.0%) | 700 | — | 1 (50.0%) | 441 | — | | | | | | 2 |
| Totals | 3 | | | 11 | | | 4 | | | 7 | | | 1 | | 26 |

stances, good results were obtained with depth doses of less than 200 r. Rarely was it found advisable to exceed 450 r (depth). If a lesion failed to respond to this amount, higher doses held no promise of success.

### HEMATURIA

Hematuria is a frequent complication of acute leukemia in childhood, particularly during the terminal phases when thrombocytopenia is marked and hemorrhage into various organs and tissues is common. It may, however, occur at an earlier stage of the illness and has been the presenting symptom of some children.[20] While the loss of blood during a twenty-four hour period is rarely of major concern, persistent hematuria can lead to a significant drop in hemoglobin levels. Extensive leukemic involvement of the kidneys, and less so of the bladder and ureters, is a common finding at autopsy.[10,11,16] In the kidneys, the changes range from scattered petechial hemorrhages in an otherwise apparently normal organ to massive nephromegaly caused by aggregates of leukemic cells infiltrating the renal substance and the epithelial surfaces. Findings in the bladder or ureters are rarely so pronounced, although infiltrates may be found in the walls of these structures.

Many of the 372 children reviewed developed hematuria at some time during the course of their illness, but only 18 received roentgen treatment (Table 1). The others were managed successfully by other methods,[12] or were in the final stage of their disease. No form of treatment, including roentgen therapy, has proved to be of consistent value during the terminal phase and irradiation is, therefore, employed infrequently at such a time. Those patients who received roentgen therapy had grossly bloody urine which had persisted for more than twenty-four hours. Although the kidneys usually show the most striking changes pathologically, it has been found empirically that roentgen therapy of the urinary bladder more often leads to satisfactory results, hematuria clearing well in 35 per cent of the instances treated (Table VIII).

Roentgen therapy is given using technique E or F. Two hundred r (air) daily is delivered for four to six days through an anterior portal to attain a depth dose of 450 r to 500 r in the bladder. However, should the hematuria clear before the projected total amount is given, or if this dose does not result in immediate benefit, treatments are discontinued. When higher doses are accumulated after initial failure, the response to roentgen therapy continues to be poor. In 3 children, the kidneys were irradiated with an average depth dose of 271 r after prior roentgen treatment of the bladder had been of no benefit. Two of

TABLE VIII

HEMATURIA

RESULTS OF ROENTGEN THERAPY OF BLADDER

| | Excellent | Good | Fair | Poor | Not Evaluated | Total |
|---|---|---|---|---|---|---|
| No. of Episodes Treated | 2 (11.1%) | 5 (27.8%) | 1 (5.6%) | 10 (55.5%) | 1 | 19 |
| Depth Dose Average (r) | 475 | 458 | 426 | 473 | 800 | |
| Range (r) | 438–511 | 402–622 | | 154–830 | | |

these patients did not improve, but in the third the urine cleared well after roentgen therapy of the kidneys was instituted. This child received a depth dose of 456 r. Another patient had roentgen treatment of the kidneys first, receiving 415 r (depth), with subsequent improvement. Hematuria recurred after a brief interval and the bladder was treated with a comparable dose, also with good results.

### LEUKEMIC MASSES

Eleven patients received roentgen therapy because of symptoms and signs referable to massive leukemic involvement of various organs other than those already discussed. All were in hematologic relapse at the time of roentgen therapy. There were 14 masses in all, with the following distribution: enlarged lymph nodes, 4; infiltration of the parotid, 3; mediastinal infiltration, 2; abdominal masses, 2; splenomegaly with pain referred to the shoulder, 1; proptosis, without increased intracranial pressure, 1; enlarged kidneys without hematuria, 1. Treatment was administered because of displacement and compression of neighboring structures by the tumors, or because of discomfort attributable to their presence. Good results were obtained in 9 of the 14 masses irradiated. Treatment was administered with techniques C, D, E, or F (Table 11). Daily doses of 25 to 200 r (air) were given to the areas involved, with the total depth dose ranging from 16 to 900 r. The effective depth dose, as before, was found to be about 450 r.

### DISCUSSION

The management of children with acute leukemia is made more difficult because of the many complications seen during the course of the disease. If left untreated, these complications are progressive in our experience. They can lead to severe disability and, on occasion, precipitate early demise. Effective management, therefore, is important in order to maintain as normal a life as possible during the period of increased survival produced by antileukemic therapy. The value of roentgen irradiation for this purpose has been known for some time. Earlier reports, concerned primarily with roentgen irradiation of large areas as a means of treatment for the acute leukemic process, contain brief mention of improvement of troublesome symptoms, particularly bone pain. Irradiation of the entire body or of the hematopoietic system as a form of treatment has been abandoned, but roentgen therapy of local areas in small dosage for control of symptoms has proved to be of distinct benefit. The mechanism by which irradiation exerts its beneficial effect remains speculative, but lysis of local aggregates of abnormal cells is believed to be the chief factor. This form of empirical treatment has been used for many

years at The Children's Hospital in Boston. No untoward effects on the hematologic status have been encountered in the patients studied. Not all the secondary difficulties of acute leukemia in childhood are helped by the use of irradiation. Most of those related to the abnormal clotting mechanism encountered late in the disease are not amenable to this form of management, but some complications which occur in children who are not in the terminal stage, regardless of whether in relapse or remission, can be controlled satisfactorily by this means. This is not to advocate indiscriminate irradiation each time a transient complaint develops. However, when a disabling and persistent symptom appears, a trial of roentgen therapy in small dosage is worthy of consideration. In children with acute leukemia, there is much to be gained and nothing lost by following such a policy.

SUMMARY

In 372 children with acute leukemia, many complications occurred which involved various organ systems, and which were progressive if untreated. Most of the complications developed during hematologic relapse, although some, particularly those involving the central nervous system, were manifest during remission. None seemed related to specific chemotherapy. For complaints not responsive to other therapy, irradiation was often used, and this report describes 95 patients so treated. The most favorable results were obtained in the treatment of central nervous system symptoms and those related to the bones and joints. Satisfactory responses also were noted in patients who had cutaneous and mucosal infiltrations and in those with a miscellaneous group of leukemic masses, while hematuria was found to clear well in some children treated by this means. It was found that small doses of roentgen rays were sufficient to produce improvement if any was to be obtained. In none of the 95 patients was any hematologic deterioration attributable to the irradiation given. The technique of roentgen treatment is described and some possible dangers discussed.

Giulio J. D'Angio, M.D.
The Children's Medical Center
300 Longwood Avenue
Boston 15, Massachusetts

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# EDITORIALS

## CHANGING CONCEPTS OF MALFUNCTION OF THE GASTROESOPHAGEAL JUNCTION IN INFANTS

ONE of the common observations in the roentgen examination of the upper gastrointestinal tract of vomiting infants is the passage of opaque material from the stomach into the distal esophagus. The question then arises as to whether this represents physiologic reflux, so-called cardioesophageal chalasia, or is a manifestation of hiatal hernia. The differentiation is thought to be of considerable importance inasmuch as the prognosis and treatment of these conditions are quite different. If a definite hiatus hernia is demonstrated, the preferred treatment usually is surgical repair, while the other two conditions may be treated medically.

Physiologic reflux may be defined as a regurgitation of gastric content into the esophagus, such as may be associated with a "burp." It is inconstant and may be seen in infants who are not vomiting despite the fact that the gastroesophageal junction is not patulous. Cardioesophageal chalasia has been defined as a hiatal incompetence with a normally situated cardia. This concept was given great impetus by Neuhauser and Berensberg[1] who reported a series of 12 cases in 1947 in which they observed persistent vomiting due to relaxation or dysfunction of the hiatal portion of the esophagus with failure of the normal "sphincter" action of the cardia. Because in many respects the condition appeared the opposite of achalasia, they chose to refer to the persistent relaxation as chalasia. Several other investigators have discussed cardioesophageal relaxation, both prior to and after Neuhauser and Berensberg's paper. Some of these authors shared the same opinions; others, however, such as Husfeldt, Thomsen, and Wamberg,[2] believed these cases to be best regarded as a sliding type of hiatal hernia without concomitant esophageal inflammatory changes. It would thus appear that the problem, at least academically, has not been solved. This would seem to be a direct result of our lack of precise knowledge of the anatomic and physiologic factors governing the function of the gastroesophageal junction.

It is generally agreed that there is a closing mechanism operative at the cardia which normally prevents regurgitation. Yet, the existence of an intrinsic anatomic sphincter has not been demonstrated to the satisfaction of most observers. Allison[3] ascribes a definite significance to the pinchcock action of the diaphragmatic reflections which form a sling about the distal esophagus and which prevent reflux during inspiration. Barrett,[4] in 1952, asserted that the oblique entry of the esophagus into the stomach alone accounts for the sphincteric action and concluded it to be present only when the acute angle between the greater curvature and the lower portion of the esophagus is maintained and controlled. A hernia by definition may be regarded as a situation in which any of the abdominal viscera extend beyond the usual confines of the peritoneal cavity. Would it not then be proper to regard the demonstration of gastric mucosa above the hiatus as a hernia? It has been shown by Palmer[5] that silver clips placed at the esophagogastric junction in normal individuals may move

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as much as 1 to 2 cm. when various maneuvers are executed during fluoroscopy. This is believed to be only a mucosal migration without attendant change in position of the muscular wall. In the adult, several anatomic landmarks such as the phrenoesophageal ligaments, inferior esophageal sphincter, and cardial ring may often be localized with a fair degree of certainty, but unfortunately they cannot be delineated nearly as well in the infant.

The roentgen examination is carried out with two purposes in mind. The first is to attempt to demonstrate or produce the reflux phenomenon and the second is to study the mucosal pattern in an effort to locate the esophagogastric junction, if possible. A rather thick barium paste is given and the stomach is distended either as a result of the simultaneously ingested air or, if necessary, by the administration of additional thin barium. The thick barium mixture coats the wall of the esophagus, provides good distention, and also enables the study of the mucosal pattern after the main bolus has entered the stomach. The normal esophagus has two to four fine linear folds; the gastric mucosa is more irregular and coarse, although in some cases the gastric folds may also appear to be linear and may present as close streaks varying from four to ten in number. The various maneuvers used are designed to produce reflux of the stomach content into the distal esophagus. The lumen of the esophagus usually appears larger and the hiatus at one time or other is widely patent. The reflux may be most pronounced during inspiration. Dornhorst, Harrison, and Pierce[6] by recording simultaneous pressures in the esophagus and stomach found a functional cardiac "valve" of about 5 mm. length at the esophagogastric junction, which offered a resistance to the forward flow of 4 mm. of mercury. During inspiration, the difference in pressure was 15 mm., a fact which favors reflux. The Müller maneuver created a difference of 80 mm. pressure. In the normal adult there was no tendency to pressure equalization, indicating a competent cardia. Creamer[7] showed that in some patients with hiatus hernia the usual inverse relationship was not present and thought that this provided a basis for incompetence. All hernias, however, are not associated with this phenomenon and, as would be expected, the reflux may be inconstant or completely absent. Hence, other maneuvers may be necessary to raise the gastric or intra-abdominal pressure. Among these are overdistention of the stomach by gas, fluid, or both; manual pressure applied directly to the epigastrium; extreme Trendelenburg positioning; or simply the struggling of the infant while being examined. To be significant, the reflux which is produced must be reproducible or constant.

The size of the hiatus may be important, although it is stated by Barrett[4] that normal hiatal size is hard to define. A better way may be to determine how long the gastroesophageal lumen remains open. The angle between the esophagus and stomach is estimated with the thought that a more centrally located esophagus is more apt to exhibit the reflux phenomenon.

Of what significance is reflux in relationship to hiatus hernia? The essential factor may be the presence, or absence, of a persistent complicating esophagitis. This may be difficult to determine roentgenologically unless the changes become marked and involve more than the mucosa. Intermittent spasm, mucosal irregularity, frank ulceration, or even stricture in an advanced case may be helpful signs. Such cases should be followed closely and treated vigorously, although the less obvious ones warrant the same vigorous regimen, since 10 per cent of them ultimately develop strictures, and the cessation of vomiting might not be due to a cure but rather to stricture formation. The role played by hormonal factors, excessive gastric acidity, and an ectopic gastric mucosa may also be profitably studied in relationship to the problem of reflux. A knowledge of the fate of these individuals upon reaching adult life could greatly aid in devising the best approach to the treatment of this condition in infancy.

JOSEPH O. REED, M.D.
Children's Hospital of Michigan
Detroit, Michigan

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# IN MEMORIAM

DR. JAMES NEWTON COLLINS of Indianapolis, Indiana died January 3, 1959 of a coronary artery occlusion.

Jim Collins was born in Fayette, Alabama in 1895. He received his A.B. degree from the University of Alabama and his M.D. from Emory University Medical School. After his internship, he decided to specialize in radiology and studied at the University of Michigan while Dr. Preston M. Hickey was Chief of the Department of Radiology. He remained there for two years and then went to Indianapolis where he joined the group of Drs. Cole, Beeler and Smith. He became well known in the field of radiation therapy. He was on the staff of St. Vincent's and Methodist Hospitals and was Associate Professor, Department of Radiology, Indiana University School of Medicine.

Doctor Collins was active in all medical circles. He was a member of his county and state societies and the A.M.A. He was an active member of the Radiological Society of North America and served several years on the Scientific Exhibit Committee. He was a member of the American Roentgen Ray Society and active for many years as Secretary-Treasurer of the Indiana Roentgen Society.

Doctor Collins is survived by his wife, Josephine Washburn Collins; a daughter, Jane Collins Wilkerson; and a son, Steven W. He also had one grandson, Bruce. He was a member of the Second Presbyterian Church.

Doctor Collins was exceptionally well grounded in the physical aspects of his specialty. He was the author of several medical papers, most of which dealt with the subject of radiation therapy, a field in which he was well-known and respected throughout Indiana. Radiology, particularly the private practice of radiology, has lost a valuable servant.

RAYMOND C. BEELER, M.D.

# SOCIETY PROCEEDINGS AND NEWS ITEMS

## MEETINGS OF RADIOLOGICAL SOCIETIES*

### UNITED STATES OF AMERICA

**AMERICAN ROENTGEN RAY SOCIETY**
*Secretary*, Dr. C. Allen Good, Mayo Clinic, Rochester, Minn. Annual meeting: Netherland-Hilton Hotel, Cincinnati, Ohio, Sept. 22–25, 1959.

**AMERICAN RADIUM SOCIETY**
*Secretary*, Dr. Robert L. Brown, Winship Clinic, Emory University, Atlanta 22, Ga. Annual meeting: Caribe Hilton, San Juan, Puerto Rico, March 17–19, 1960.

**RADIOLOGICAL SOCIETY OF NORTH AMERICA**
*Secretary*, Dr. D. S. Childs, 713 E. Genesee St., Syracuse 2, N. Y. Annual meeting: Palmer House, Chicago, Ill., Nov. 15–20, 1959.

**AMERICAN COLLEGE OF RADIOLOGY**
*Executive Director*, William C. Stronach, 20 N. Wacker Drive, Chicago 6, Illinois. Annual meeting: Feb. 3–6, 1960, Roosevelt Hotel, New Orleans, La.

**SECTION ON RADIOLOGY, AMERICAN MEDICAL ASSOCIATION**
*Secretary*, Dr. T. Leucutia, Harper Hospital, Detroit 1, Mich. Annual meeting: June 13–17, 1960, Miami Beach, Fla.

**AMERICAN BOARD OF RADIOLOGY**
*Secretary*, Dr. H. Dabney Kerr. Correspondence should be directed to Kahler Hotel Building. Rochester, Minn. The Board will conduct a Special Examination in Nuclear Medicine (for diplomates in Radiology or Therapeutic Radiology) on Dec. 5, 1959 at the Shoreham Hotel in Washington, D. C. Deadline for filing applications was Aug. 1, 1959. The regular fall examination will be held at the Shoreham Hotel, December 6–9, 1959. Deadline for filing applications was July 1, 1959. Candidates examined in Diagnostic Roentgenology may expect to be examined in Physics.

**NINTH INTERNATIONAL CONGRESS OF RADIOLOGY**
*Secretary-General*, Dr. Hans v. Braunbehrens. Request for information should be addressed to Dr. Viktor Loeck, Kongressbüro, Forsthausstrasse 76, Frankfurt am Main, Germany. Met in Munich, Germany, July 23 to July 30, 1959. Next meeting will be announced.

**SEVENTH INTER-AMERICAN CONGRESS OF RADIOLOGY**
*Secretary*, Dr. Laura Fariñas, Havana, Cuba. *Counselor for the United States*, Dr. J. A. del Regato, Penrose Cancer Hospital, 2200 North Cascade Avenue, Colorado Springs, Colorado. The meeting will be held in Brazil in 1961.

**ALABAMA RADIOLOGICAL SOCIETY**
*Secretary*, Dr. J. A. Meadows, Jr., Medical Arts Bldg., Birmingham 5, Ala. Meets time and place Alabama State Medical Association.

**AMERICAN NUCLEAR SOCIETY**
*Executive-Secretary*, Octave J. Du Temple, 86 E. Randolph St., Chicago, Ill.

**ARIZONA RADIOLOGICAL SOCIETY**
*Secretary*, Dr. R. Lee Foster, 1313 N. Second St., Phoenix, Ariz. Two regular meetings a year. Annual meeting at time and place of State Medical Association and interim meeting six months later.

**ARKANSAS RADIOLOGICAL SOCIETY**
*Secretary*, Dr. J. B. Scruggs, Arkansas Baptist Hospital, Little Rock, Ark. Meets every three months and also at time and place of State Medical Association.

**ASSOCIATION OF UNIVERSITY RADIOLOGISTS**
*Secretary*, Dr. Paul Riemenschneider, Medical College State University of New York, Syracuse, N. Y. Annual meeting to be announced.

**ATLANTA RADIOLOGICAL SOCIETY**
*Secretary*, Dr. J. L. Clements, Jr., 35 Linden Ave., N.E., Atlanta 8, Ga. Meets monthly, except during three summer months, on second Friday evening.

**BLOCKLY RADIOLOGICAL SOCIETY**
*Secretary*, Dr. Samuel Finkelman, 101 S. Twentieth St. Philadelphia, Pa.

**BROOKLYN RADIOLOGICAL SOCIETY**
*Secretary*, Dr. George A. Manfredonia, One Hanson Place, Brooklyn, N. Y. Meets first Thursday of each month October through May.

**BUFFALO RADIOLOGICAL SOCIETY**
*Secretary*, Dr. Alfred Dobrak, 710 Leydecker Rd., Buffalo 24, N. Y. Meets second Monday evening each month, October to May inclusive.

**CENTRAL NEW YORK RADIOLOGICAL SOCIETY**
*Secretary*, Dr. Joseph A. Head, 150 Marshall St., Syracuse, N. Y. Meets first Monday each month October through May.

**CENTRAL OHIO RADIOLOGICAL SOCIETY**
*Secretary*, Dr. William B. Schwartz, 1500 W. Third Ave., Columbus, Ohio. Meets at 6:30 P.M. on second Thursday of October, November, January, March and May at Fort Hayes Hotel, Columbus, Ohio.

**CENTRAL SOCIETY OF NUCLEAR MEDICINE**
*Secretary*, Dr. Robert S. Landauer, Radiation Center Building, 1903 West Harrison St., Chicago 12, Ill.

**CHICAGO ROENTGEN SOCIETY**
*Secretary*, Dr. George B. Cahill, 802 Burns Ave., Flossmoor, Ill. Meets second Thursday of each month, October to April except December at the Sheraton Hotel at 8:00 P.M.

**CLEVELAND RADIOLOGICAL SOCIETY**
*Secretary*, Dr. Frederick A. Rose, 2065 Adelbert Rd., Cleveland 6, Ohio. Meetings at 7:00 P.M. on fourth Monday of each month from October to April at Tudor Arms Hotel.

**COLORADO RADIOLOGICAL SOCIETY**
*Secretary*, Dr. Lorenz R. Wurtzebach, 601 E. Nineteenth Ave., Denver, Colo. Meets third Friday of each month at Denver Athletic Club.

**CONNECTICUT VALLEY RADIOLOGIC SOCIETY**
*Secretary*, Dr. Paul J. Kingston, 114 Woodland St., Hartford, Conn. Meets first Friday in February and April.

**DALLAS-FORT WORTH RADIOLOGICAL CLUB**
*Secretary*, Dr. A. H. Keene, 3707 Gaston Ave., Dallas, Texas. Meets monthly, third Monday, at Greater Fort Worth International Airport at 6:30 P.M.

**DETROIT ROENTGEN RAY AND RADIUM SOCIETY**
*Secretary*, Dr. Kenneth L. Krabbenhoft, Harper Hospital, Detroit 1, Mich. Meets monthly first Thursday, October through May, at David Whitney House, 1010 Antietam, at 6:30 P.M.

**EAST BAY ROENTGEN SOCIETY**
*Secretary*, Dr. Dan Tucker, 434 30th St., Oakland 9, Calif. Meets first Thursday each month at Peralta Hospital, Oakland.

**EAST TENNESSEE RADIOLOGICAL SOCIETY**
*Secretary*, Dr. Charles W. Reavis, Baroness Erlanger Hospital, Chattanooga, Tenn. Meets in January and September.

*** Secretaries of societies are requested to send timely information promptly to the Editor.**

Eastern Radiological Society
*Secretary*, Dr. John D. Osmond, Jr., Euclid-Glenville Hospital, Cleveland 19, Ohio. Meets at Mid Pines Club, Southern Pines, N. C., March 6–10, 1960.

Florida Radiological Society
*Secretary*, Dr. Russell D. D. Hoover, 49th St., West Palm Beach, Fla. Meets twice annually, in the spring with the annual State Society meeting, and in the fall.

Florida West Coast Radiological Society
*Secretary-Treasurer*, Dr. Joseph C. Rush, 1501 Jungle, St. Petersburg, Fla.

Georgia Radiological Society
*Secretary*, Dr. George W. Brown, Griffin, Ga. Meets Spring and Fall with Annual State Society Meeting.

Greater Miami Radiological Society
*Secretary*, Dr. George P. Daurelle, Jackson Memorial Hospital, Miami 36, Fla. Meets monthly third Wednesday at 8 P.M. at Jackson Memorial Hospital, Miami, Fla.

Greater St. Louis Radiological Society
*Secretary*, Dr. Harvey A. Humphrey, 462 N. Taylor, St. Louis 8, Mo.

Houston Radiological Society
*Secretary*, Dr. Edward B. Singleton, 6621 Fannin St., Houston 25, Texas. Meets last Monday each month, Seminar Room, Doctors' Club of Houston.

Idaho State Radiological Society
*Secretary*, Dr. Charles R. McWilliams, Magic Valley Memorial Hospital, Twin Falls, Idaho. Meets annually at same time as Idaho State Medical Association.

Illinois Radiological Society
*Secretary*, Dr. Stephen L. Casper, 1101 Maine St., Quincy, Ill. Meets three times a year.

Indiana Roentgen Society
*Secretary*, Dr. David E. Wheeler, 1500 North Ritter, Indianapolis, Ind. Meets first Sunday in May and during fall meeting of Indiana State Medical Association.

Iowa Radiological Society
*Secretary*, Dr. James T. McMillan, 1104 Bankers Trust Bldg., Des Moines, Iowa. Luncheon and business meeting during annual session of Iowa State Medical Society. The scientific section is held in the autumn.

Kansas Radiological Society
*Secretary*, Dr. Lewis G. Allen, 807 Huron Bldg., Kansas City, Kansas. Meets in spring with State Medical Society, and in winter on call.

Kentucky Radiological Society
*Secretary*, Dr. Robert H. Akers, V. A. Hospital, Louisville 2, Ky. Meets monthly on second Friday at Seelbach Hotel, Louisville.

Kings County Radiological Society
*Secretary*, Dr. C. P. Naidorf, 411 Parkside Ave. Brooklyn 26, N. Y. Meets Kings County Med. Soc. Bldg. monthly on fourth Thursday, October to May, 8:45 P.M.

Los Angeles Radiological Society
*Secretary*, Dr. Robert B. Engle, St. Luke Hospital, Pasadena, Calif. Meets second Wednesday of month in September, November, March, April and June at Los Angeles County Medical Association Building, Los Angeles.

Maine Radiological Society
*Secretary*, Dr. Francis J. O'Connor, Augusta General Hospital, Augusta, Maine. Meets in June, September, December and March.

Maryland Radiological Society
*Secretary*, Dr. Nathan B. Hyman, 1805 Eutaw Place, Baltimore 17, Md.

Memphis Roentgen Society
*Secretary*, Dr. Hollis H. Halford, Kennedy V.A. Hospital, Department of Radiology, Memphis 15, Tenn. Meets first Monday of each month at John Gaston Hospital.

Miami Valley Radiological Society
*Secretary*, Dr. S. F. Johnson, 2197 Los Arrow Dr., Dayton 9, Ohio. Meets second Friday of fall and winter months.

Mid-Hudson Radiological Society
*Secretary*, Dr. Joseph Sorrentino, St. Francis Hospital, Poughkeepsie, N. Y. Meets 8:30 P.M., fourth Wednesday each month, September to May.

Milwaukee Roentgen Ray Society
*Secretary*, Dr. Joseph F. Wepfer, 5000 W. Chambers St., Milwaukee 10, Wis. Meets monthly on fourth Monday at University Club.

Minnesota Radiological Society
*Secretary*, Dr. Donald H. Peterson, 853 Medical Arts Bldg., Minneapolis 2, Minn. Meets three times annually, in fall, winter and spring.

Mississippi Radiological Society
*Secretary*, Dr. Bernard T. Hickman, University Medical Center, Jackson, Miss. Meets third Thursday of each month at Hotel Edwards, Jackson, at 6:00 P.M.

Montana Radiological Society
*Secretary*, Dr. Arthur T. Austin, 104 The Doctors Bldg., Billings, Montana. Meets at least once a year.

Nassau Radiological Society
*Secretary*, Dr. Jerome Zwanger, 126 Hicksville Rd., Massapequa, L. I., N. Y. Meets second Tuesday of the month in February, April, June, October and December.

Nebraska Radiological Society
*Secretary*, Dr. Warren Q. Bradley, 924 Sharp Bldg., Lincoln, Nebraska. Meets third Wednesday of each month at 6 P.M. in Omaha or Lincoln.

New England Roentgen Ray Society
*Secretary*, Dr. John E. Gary, 1180 Beacon St., Brookline 46, Mass. Meets third Friday of each month, October through May, at The Longwood Towers, Brookline, Mass.

New Hampshire Roentgen Ray Society
*Secretary*, Dr. Paul Y. Hasserjian, 1470 Elm St., Manchester, N. H. Meets four to six times yearly.

New York Roentgen Society
*Secretary*, Dr. William B. Seaman, 622 W. 168th St., New York, N. Y. Meets monthly on third Monday, New York Academy of Medicine, at 4:30 P.M.

North Carolina Radiological Society
*Secretary*, Dr. Charles A. Bream, 211 McCauley St., Chapel Hill, N. C. Meets in May and October.

North Dakota Radiological Society
*Secretary*, Dr. Marianne Wallis, St. Joseph's Hospital, Minot, N. D. Meetings by announcement.

North Florida Radiological Society
*Secretary*, Dr. Paul A. Mori, 800 Miami Road, Jacksonville 7, Fla. Meets quarterly in March, June, September and December.

Northeastern New York Radiological Society
*Secretary*, Dr. Irving Van Woert, Jr., Albany Hospital, Albany, N. Y. Meets in Albany area on second Wednesday of October, November, March and April.

Northern California Radiological Society
*Secretary*, Dr. Rob H. Kirkpatrick, 1219 28th St., Sacramento, Calif. Meets at dinner last Monday of each month, September to June.

Ohio State Radiological Society
*Secretary*, Dr. Paul D. Meyer, 125 S. Grant Ave., Columbus, Ohio. Annual meeting: Cincinnati, Ohio, May, 1959.

Oklahoma State Radiological Society
*Secretary*, Dr. J. Murphree, 617 E. Brookfield, Ponca City, Okla. Three regular meetings annually.

Oregon Radiological Society
*Secretary*, Dr. C. V. Allen, Veterans Hospital, Portland, Ore. Meets monthly from October to June on the second Wednesday of each month at 8:00 P.M. at the University Club.

ORLEANS PARISH RADIOLOGICAL SOCIETY
*Secretary*, Dr. Joseph V. Schlosser, Charity Hospital, New Orleans 13, La. Meets second Tuesday of each month.

PACIFIC NORTHWEST RADIOLOGICAL SOCIETY
*Secretary*, Dr. J. Arch Colbrunn, 19 Royal Oak Drive, Roseburg, Ore. Annual meeting: Vancouver, British Columbia, May, 1960.

PACIFIC ROENTGEN SOCIETY
*Secretary*, Dr. L. H. Garland, 450 Sutter St., San Francisco 8, Calif. Meets annually during meeting of California Medical Association.

PENNSYLVANIA RADIOLOGICAL SOCIETY
*Secretary*, Dr. Walter P. Bitner, 234 State St., Harrisburg, Pa.

PHILADELPHIA ROENTGEN RAY SOCIETY
*Secretary*, Dr. Robert B. Funch, Department of Radiology, Germantown Hospital, Philadelphia 44, Pa. Meets first Thursday of each month, at 5 P.M., from October to May in Thompson Hall, College of Physicians.

PITTSBURGH ROENTGEN SOCIETY
*Secretary*, Dr. Charles N. Chasler, 3500 Fifth Ave., Pittsburgh 13, Pa. Meets second Wednesday of month, October through June at Park Schenley Restaurant.

RADIOLOGICAL SECTION, BALTIMORE MEDICAL SOCIETY
*Secretary*, Dr. James K. V. Willson, 1100 N. Charles St., Baltimore 1, Md. Meets third Tuesday each month, September to May, inclusive.

RADIOLOGICAL SOCIETY OF GREATER CINCINNATI
*Secretary*, Dr. John E. Singer, 468 Doctors' Bldg., Cincinnati 2, Ohio. Meets monthly from September to May on first Monday of each month at 7:30 P.M. at the Cincinnati General Hospital.

RADIOLOGICAL SOCIETY OF HAWAII
*Secretary*, Dr. Louis L. Buzaid, The Queen's Hospital, Box 861, Honolulu, Hawaii. Meets third Monday of each month at 7:30 P.M.

RADIOLOGICAL SOCIETY OF GREATER KANSAS CITY
*Secretary*, Dr. Samuel B. Chapman, Jr., 830 Argyle Bldg., Kansas City, Mo. Meets last Friday of each month.

RADIOLOGICAL SOCIETY OF KANSAS CITY
*Secretary*, Dr. Arthur B. Smith, 800 Argyle Bldg., Kansas City, Mo. Meets third Thursday of each month.

RADIOLOGICAL SOCIETY OF LOUISIANA
*Secretary*, Dr. Seymour Ochsner, Ochsner Clinic, New Orleans 15, La. Meets annually during Louisiana State Medical Society meeting.

RADIOLOGICAL SOCIETY OF NEW JERSEY
*Secretary*, Dr. Austin J. Tidaback, 912 Prospect Ave., Plainfield, N. J. Meets at Atlantic City at time of State Medical Society and in November in Newark, N. J.

RADIOLOGICAL SOCIETY OF NEW YORK STATE
*Secretary-Treasurer*, Dr Mario C. Gian, 610 Niagara St., Buffalo 1, N. Y. Annual meeting: To be announced.

RADIOLOGICAL SOCIETY OF SOUTH DAKOTA
*Secretary-Treasurer*, Dr. Donald J. Peik, 303 S. Minnesota Ave., Sioux Falls, S. D.

RADIOLOGICAL SOCIETY OF SOUTHERN CALIFORNIA
*Secretary*, Dr. Joseph F. Linsman, 436 N. Roxbury Dr., Beverly Hills, Calif.

REDWOOD EMPIRE RADIOLOGICAL SOCIETY
*Secretary*, Dr. Lee E. Titus, 164 W. Napa St., Sonoma, Calif. Meets second Monday every other month.

RICHMOND COUNTY RADIOLOGICAL SOCIETY
*Secretary*, Dr. W. F. Hamilton, Jr., University Hospital, Augusta, Ga. Meets first Thursday of each month at various hospitals.

ROCHESTER ROENTGEN RAY SOCIETY, ROCHESTER, N. Y.
*Secretary*, Dr. Theodore F. Van Zandt, 501 W. Main St., Rochester 8, N. Y. Meets at 8:15 P.M. on the last Monday of each month, September through May, at Strong Memorial Hospital.

ROCKY MOUNTAIN RADIOLOGICAL SOCIETY
*Secretary*, Dr. John H. Freed, 4200 East Ninth Ave., Denver 20, Colo. Annual meeting: Shirley-Savoy Hotel, Denver, Colorado, Aug. 20–22, 1959.

SAN ANTONIO-MILITARY RADIOLOGICAL SOCIETY
*Secretary*, Dr. Hugo F. Elmendorf, Jr., 730 Medical Arts Bldg., San Antonio 5, Texas. Meets third Wednesday each month in Fort Sam Houston Officer's Club at 6:30 P.M.

SAN DIEGO RADIOLOGICAL SOCIETY
*Secretary*, Dr. Stanley A. Moore, 2466 First Ave., San Diego 1, Calif. Meets first Wednesday of each month at the University Club.

SAN FRANCISCO RADIOLOGICAL SOCIETY
*Secretary*, Dr. M. A. Sisson, 450 Sutter St., San Francisco 8, Calif. Meets quarterly at Grison's Steak House.

SECTION ON RADIOLOGY, CALIFORNIA MEDICAL ASSOCIATION
*Secretary*, Dr. William H. Graham, 630 East Santa Clara St., San Jose, Calif.

SECTION ON RADIOLOGY, CONNECTICUT STATE MEDICAL SOCIETY
*Secretary*, Dr. Ralph J. Littwin, Bristol Hospital, Bristol, Conn. Meetings are held bi-monthly.

SECTION ON RADIOLOGY, MEDICAL SOCIETY OF THE DISTRICT OF COLUMBIA
*Secretary*, Dr. Charles E. Bickham, Jr., 1835 Eye St., N.W., Washington 6, D. C. Meets Medical Society Library, third Wednesday of January, March, May and October at 8:00 P.M.

SECTION ON RADIOLOGY, ILLINOIS STATE MEDICAL SOCIETY
*Secretary*, Dr. William Meszaros, 1825 W. Harrison St., Chicago, Ill.

SECTION ON RADIOLOGY, SOUTHERN MEDICAL ASSOCIATION.
*Secretary*, Dr. Seymour Ochsner, Ochsner Clinic, 3503 Prytania St., New Orleans 15, La. Annual meeting: Nov. 16–19, 1959, Atlanta, Ga.

SHREVEPORT RADIOLOGICAL CLUB
*Secretary*, W. R. Harwell, 608 Travis St., Shreveport, La. Meets monthly on third Wednesday, at 7:30 P.M. September to May inclusive.

SOCIETY OF NUCLEAR MEDICINE
*Secretary*, Dr. Robert W. Lackey, 452 Metropolitan Bldg., Denver 2, Colo. Annual meeting to be announced.

SOUTH BAY RADIOLOGICAL SOCIETY
*Secretary*, Dr. Stanford B. Rossiter, 1111 University Dr., Menlo Park, Calif. Meets second Wednesday of each month.

SOUTH CAROLINA RADIOLOGICAL SOCIETY
*Secretary*, Dr. Wayne Reeser, 804 Burroughs St., Conway, S. C. One meeting with South Carolina Medical Association in May, others arranged by President.

SOUTHERN RADIOLOGICAL CONFERENCE
*Secretary*, Dr. Marshall Eskridge, Mobile Infirmary, Mobile, Ala.

SOUTHWESTERN RADIOLOGICAL SOCIETY
*Secretary*, Dr. Gordon Black, Suite 2-A, El Paso Medical Center, 1501 Arizona, El Paso, Texas. Meets second Tuesday of each month.

TENNESSEE RADIOLOGICAL SOCIETY
*Secretary*, Dr. James J. Range, P.O. Box 324, Johnson City, Tenn. Meets annually at the time and place of the Tennessee State Medical Association.

TEXAS RADIOLOGICAL SOCIETY
*Secretary*, Dr. R. P. O'Bannon, 402 Professional Bldg., 1216 Pennsylvania Ave., Fort Worth 4, Texas. Next meeting January 22 and 23, 1960, Houston, Texas.

TRI-STATE RADIOLOGICAL SOCIETY
*Secretary*, Dr. James R. Mathews, 118 S. E. First St., Evansville, Ind. Meets last Wednesday of Oct., Jan., March and May, 8:00 P.M. at Elks' Club in Evansville.

UNIVERSITY OF MICHIGAN DEPARTMENT OF ROENTGENOLOGY STAFF MEETING
Meets each Monday evening from September to June, at 7:00 P.M. at University Hospital.

Upper Peninsula Radiological Society
*Secretary*, Dr. A. Gonty, Menominee, Mich. Meets quarterly.

Utah State Radiological Society
*Secretary*, Dr. William R. Christensen, 2033 S. State St., Salt Lake City, Utah. Meets fourth Wednesday in January, March, May, September and November at Holy Cross Hospital.

Virginia Radiological Society
*Secretary*, Dr. Frank A. Kearney, II, 110 S. Curry St., Phoebus, Va. Meets annually in October.

Washington State Radiological Society
*Secretary*, Dr. Wayne A. Chesledon, 306 Stimson Bldg., Seattle 1, Wash. Meets fourth Monday of each month from September through May at 610 Pine St., Seattle.

West Virginia Radiological Society
*Secretary*, Dr. Karl J. Myers, 112 N. Woods St., Philippi, W. Va. Meets concurrently with annual meeting of West Virginia State Medical Society; other meetings arranged by program committee.

Westchester Radiological Society
*Secretary*, Dr. Arnold Myron Wald, 406 Boston Post Road, Port Chester, N. Y. Meets on third Tuesday of January and October and on two other dates.

Wisconsin Radiological Society
*Secretary*, Dr. Farrell F. Golden, 5221 Tonyawatha Trail, Madison 4, Wis. Special meetings held during year; annual meeting each spring at Madison, Wis.

X-Ray Study Club of San Francisco
*Secretary*, Dr. John H. Heald, 450 Sutter St., San Francisco 8, Calif. Meets monthly, third Thursday at 7:30 P.M., Children's Hospital, September through June.

### Cuba, Mexico, Puerto Rico and Central America

Asociación de Radiólogos de Centro America y Panamá
Comprising: Guatemala, El Salvador, Honduras, Nicaragua, Costa Rica and Panamá, *Secretary-General*, Dr. Roberto Calderón, Calle Central Deste No. 218, Managua, Nicaragua. Meets annually in a rotating manner in the six countries.

Sociedad de Radiología de El Salvador
*Secretary*, Dr. Rafael Vega Gómez.

Sociedad de Radiología de Guatemala
*Secretary*, Dr. Carlos E. Escobar, 9ª. Calle A 0-05, Zona 1, Guatemala.

Sociedad de Radiología y Fisioterapía Cubana
*Secretary*, Dr. Miguel A. García Plasencia, Hospital Curie, 29 y F, Vedado, Habana, Cuba. Meets monthly at Curie Hospital.

Sociedad Costarricense de Radiologia
*Secretary*, Dr. James Fernández Carballo, Apartado VIII, San José, Costa Rica.

Sociedad Mexicana de Radiología, A. C.
Calle del Oro No. 15, México 7, D. F.
*Secretary-General*, Dr. Guillermo Santín. Meets first Monday of each month.

Asociación Puertorriqueña de Radiología
*Secretary*, Dr. R. B. Díaz Bonnet, Suite 504, Professional Bldg., Santurce, Puerto Rico.

Sociedad Radiológica Panameña
*Secretary*, Dr. L. Arrieta Sánchez, Apartado No. 86, Panamá, R. de P. Meets monthly in a department of radiology of a local hospital, chosen at preceding meeting.

### British Commonwealth of Nations

Association of Radiologists of the Province of Quebec
*Secretary*, Dr. Odilon Raymond, 5400 Blvd. Gouin. Quest, Montreal, Que. Meets four times a year.

British Institute of Radiology Incorporated with the Röntgen Society
*Honorary Secretary*, Dr. John Blewett, 32 Welbeck St., London, W. 1. Meets monthly from October until May.

Faculty of Radiologists
*Honorary Secretary*, Dr. R. A. Kemp Harper, 47 Lincoln's Inn Fields, London, W.C.2, England.

Section of Radiology of the Royal Society of Medicine (Confined to Medical Members)
Meets third Friday each month at 4:45 P.M. at the Royal Society of Medicine, 1 Wimpole St., London, W. 1.

Canadian Association of Radiologists
*Honorary Secretary*, Dr. Robert G. Fraser, *Associate Honorary Secretary*, Dr. Jean-Louis Léger, 1555 Summerhill Ave., Montreal 25, Que. Meetings January and June.

Montreal Radiological Study Club
*Secretary*, Douglas W. MacEwan, 2300 Tupper St., Montreal 25, Quebec. Meets first Tuesday evening, October to April.

Section of Radiology, Canadian Medical Association
*Secretary*, Dr. C. M. Jones, Inglis St., Ext. Halifax, N. S.

Société Canadienne-Française d'Electro-Radiologie Médicale
*General Secretary*, Dr. Louis Ivan Vallée, 1058 rue St-Denis, Montreal 18, Canada. Meets third Saturday each month.

Toronto Radiological Society
*Secretary*, Dr. L. R. Harnick, Toronto Western Hospital, 399 Bathurst St., Toronto, Ontario. Meets second Monday of each month September through May.

College of Radiologists of Australasia
*Honorary Secretary*, Dr. E. A. Booth, c/o British Medical Agency, 135 Macquarie St., Sydney, N.S.W., Australia.

### South America

Asociación Argentina de Radiología
*Secretary*, Dr. Lidio G. Mosca, Avda. Gral. Paz 151, Córdoba, Argentina. Meetings held monthly.

Ateneo de Radiologia
*Secretary*, Dr. Victor A. Añaños, Instituto de Radiologia, Santa Fe 3100, Rosario, Argentina. Meets monthly on second and fourth Fridays at 7:00 P.M. in the Hospital Nacional del Centenario, Santa Fe 1300, Rosario.

O Colégio Brasileiro de Radiologia
*Secretary-General*, Dr. Walter Bomfins Pontes. Seat: Rua Major Quedinho, 99-2°. Andar, São Paulo, Brazil.

Sociedad Argentina de Radiología, Junta Central, Buenos Aires
*Secretary*, Dr. Edgardo O. Olcese, Santa Fé 1171, Buenos Aires. Meetings are held monthly.

Sociedad Bolivana de Radiología
*Secretary*, Dr. Javier Prada Méndez, Casilla 1596, La Paz, Bolivia. Meets monthly. General assembly once every two years.

Sociedade Brasileira de Radiologia
*Secretary*, Dr. Nicola Caminha, Av. Mem. de Sa, Rio de Janeiro, Brazil. General Assembly meets every two years in December.

Sociedade Brasileira de Radioterapia
*Secretary*, Dr. Oscar Rocha von Pfuhl, Av. Brigadeiro Luiz Antonio, 644 São Paulo, Brazil. Meets monthly on second Wednesday at 9:00 P.M. in São Paulo at Av. Brigadeiro Luiz Antonio, 644.

Sociedad Chilena de Radiología
*Secretary*, Dr. Juan Sabbagh Dacla, Merced 565, Santiago, Chile. Meets fourth Friday of each month.

Sociedad Colombiana de Radiologia
*Secretary*, Dr. Alberto Mejía Diazgranados, Carrera 13, No. 25–31, Apartado aéreo No. 5804, Bogotá, Colombia. Meets last Thursday of each month.

Sociedad Ecuatoriana de Radiología y Fisioterapía
*Secretary*, Dr. Publio Vargas P., Casilla 1242 Guayaquil, Ecuador.

Sociedad Paraguaya de Radiología
*Secretary*, Dr. Miguel González Addone, 15 de Agosto 322, Asunción, Paraguay.

SOCIEDAD PERUANA DE RADIOLOGIA
*Secretary*, Dr. Luis Pinillos Ganoza, Apartado 2306, Lima, Perú. Meets monthly except during January, February and March, at Asociación Médica Peruana "Daniel A. Carrión," Villalta 218, Lima.

SOCIEDAD DE RADIOLOGIA DEL ATLANTICO
*Secretary*, Dr. Raul Fernandez, Calle 40 #41-110, Baranquilla, Colombia. Society meets monthly at the Instituto de Radiologia.

SOCIEDAD DE RADIOLOGÍA, CANCEROLOGÍA Y FÍSICA MÉDICA DEL URUGUAY
*Secretary-General*, Dr. Ricardo Parada, Av. Agraciada 1464, piso B, Montevideo, Uruguay.

SOCIEDADE DE RADIOLOGIA DE PERNAMBUCO
*Secretary*, Dr. Manoel Medeiros, Instituto de Radiologia da Faculdade de Medicina da Universidade do Recife, Caixa Postal 505, Pernambuco, Brazil.

SOCIEDAD VENEZOLANA DE RADIOLOGÍA
*Secretary-General*, Dr. Rubén Merenfeld, Apartado No. 9362, Candelaria, Caracas, Venezuela. Meets monthly third Friday at Colegio Médico del Distrito Federal, Caracas.

### CONTINENTAL EUROPE

ÖSTERREICHISCHE RÖNTGEN-GESELLSCHAFT
*President*, Dr. Konrad Weiss, Mariannengasse 10, Vienna 9, Austria. Meets second Tuesday of each month in Allgemeine Poliklinik.

SOCIÉTÉ BELGE DE RADIOLOGIE
*General Secretary*, Dr. S. Masy, 256 Chaussée de Wavre, Heverlee-lez-Louvain, Belgium. Meets in February, March, May, June, October, November and December.

SOCIÉTÉ FRANÇAISE D'ELECTRORADIOLOGIE MÉDICALE, and its branches: SOCIÉTÉ DU SUD-OUEST, DU LITTORAL MÉDITERRANÉEN, DU CENTRE ET DU LYONNAIS, DU NORD, DE L'OUEST, DE L'EST, ET D'ALGER ET D'AFRIQUE DU NORD. Central Society meets third Monday of each month, except during July, August and September, rue de Seine 12, Paris.
*Secretary-General*, Dr. Ch. Proux, 9, rue Daru, Paris 8e, France.

CESKOSLOVENSK SPOLEČNOST PRO RÖNTGENOLOGII A RADIOLOGII V PRAZE
*Secretary*, Dr. Roman Bláha, Praha XII, stát. nemocnice, Czechoslovakia. Meets monthly except during July, August, and September. Annual general meeting.

DEUTSCHE RÖNTGENGESELLSCHAFT
*Secretary*, Professor Dr. med. H. Lossen, Universitäts-Röntgeninstitut. Langenbeckstr. 1, Mainz, Germany. Meets annually.

SOCIETÀ ITALIANA DI RADIOLOGIA MEDICA E DI MEDICINA NUCLEARE
*Secretary*, Dr. Ettore Conte, Ospedale Mauriziano, Torino, Italy. Meets annually.

NEDERLANDSE VERENIGING VOOR ELECTROLOGIE EN RÖNTGENOLOGIE
*Secretary*, Dr. J. R. von Ronnen, Violenweg 14, den Haag, Netherlands.

SCANDINAVIAN ROENTGEN SOCIETIES
The Scandinavian roentgen societies have formed a joint association called the Northern Association for Medical Radiology, meeting every second year in the different countries belonging to the Association.

SOCIEDAD ESPAÑOLA DE RADIOLOGIA Y ELECTROLOGIA
*Secretary*, Dr. D. Aureo Gutierrez Churruca, Esparteros, No. 9, Madrid, Spain. Meets monthly in Madrid.

SCHWEIZERISCHE GESELLSCHAFT FÜR RADIOLOGIE UND NUKLEARMEDIZIN (SOCIÉTÉ SUISSE DE RADIOLOGIE ET DE MÉDECINE NUCLÉAIRE)
*Secretary*, Dr. Max Hopf, Effingerstrasse 47, Bern, Switzerland.

### INDIA

INDIAN RADIOLOGICAL ASSOCIATION
*Secretary*, Dr. R. F. Sethna, Navsari Building, Hornby Road, Bombay 1, India.

The next list of Meetings of Radiological Societies will be published in the November issue of the JOURNAL.

## AMERICAN BOARD OF RADIOLOGY

The Board of Trustees of The American Board of Radiology has decided to postpone the obligatory examination in Nuclear Medicine until after June 30, 1962 for those candidates being examined in Radiology or Therapeutic Radiology. Up until that time, therefore, the examination in Nuclear Medicine will be optional for those who wish to be examined in Radiology or Therapeutic Radiology.

## CONTINUATION COURSE IN GASTROINTESTINAL RADIOGRAPHY FOR RADIOLOGISTS

The University of Minnesota announces a continuation course in Gastrointestinal Radiography for Radiologists from November 2 to 6, 1959. The course will be held in the Mayo Memorial Auditorium at the University of Minnesota Medical Center on the Minneapolis Campus. Guest participants will include Doctors John R. Amberg, Assistant Professor of Radiology, Marquette University School of Medicine, Milwaukee; Edward B. Benedict, Assistant Clinical Professor of Surgery, Harvard Medical School, Boston; Frank L. Campeti, Assistant Professor of Radiology, University of Rochester School of Medicine and Dentistry, Rochester, New York; Roberto L. Estrada, Surgeon, Montreal General Hospital, Montreal, Quebec; Cesare Gianturco, Director of Radiology, Carle Hospital Clinic, Urbana, Illinois; Ross Golden, Professor Emeritus of Radiology, Columbia University College of Physicians and Surgeons, New York City, and Visiting Professor of Radiology, University of California School of Medicine at Los Angeles, Los Angeles; Elliott C. Lasser, Professor and Chairman of Radiology, University of Pittsburgh School of Medicine, Pittsburgh; Richard H. Marshak, Associate Radiologist, Mount Sinai Hospital, New York City; Harry Z. Mellins, Professor and Chairman of Radiology, State University of New York College of Medicine, Brooklyn; Everett L. Pirkey, Professor and Chairman of Radiology, University of Louisville School of Medicine, Louisville; M. H. Poppel, Professor and Chairman of Radiology, New York University College of Medicine, New York City; Leo G. Rigler, Visiting Professor of Radiology, University of California School of Medicine at Los Angeles, Los Angeles; J. Maurice Robinson, Assistant Clinical Professor of Radiology, University of California School of Medicine and Stanford University School of Medicine, San Francisco; Richard Schatzki, Chief of Radiology, Mount Auburn Hospital, Cambridge, and Clinical Associate of Radiology, Harvard Medical School, Boston; Robert Shapiro, Chairman of Radiology, Hospital of St. Raphael, New Haven, Connecticut; Frederic E. Templeton, Clinical Professor of Radiology, University of Washington School of Medicine, Seattle; Solve Welin, Radiologist, Allmanaa Sjukhuset, Malmo, Sweden; and Robert E. Wise, Radiologist, Lahey Clinic and New England Baptist Hospital, Boston.

The course will be presented under the direction of Doctors Harold O. Peterson, Professor and Head, Department of Radiology; Joseph Jorgens, Associate Professor, Department of Radiology, and Chief, Radiology Service, Veterans Administration Hospital; and Dr. Norman Blank, Instructor, Department of Radiology. The remainder of the faculty will include members of the faculty of the University of Minnesota Medical School and the Mayo Foundation.

Lodging and meal accommodations are available at the Center for Continuation Study.

## SECOND ANNUAL SYMPOSIUM ON CINEFLUOROGRAPHY

The Second Annual Symposium on Cinefluorography will be held Friday and Saturday, November 13 and 14, 1959, at the University of Rochester Medical Center. This Second Symposium will emphasize the clinical and investigative value of cineangiocardiography. In addition, time will

be provided for the presentation of information on image intensifier equipment, photographic technique and apparatus, and papers dealing with other clinical uses of cinefluorography.

The program will begin Friday morning with the "Second Annual Lecture on Cinefluorography of the University of Rochester School of Medicine and Dentistry" by Dr. Frank L. Campeti. He will summarize six years of clinical and experimental work on cineangiocardiography, and will review the indications and limitations of the method. Following this, a cardiologic team will discuss the diagnosis, prognosis and surgical treatment of congenital and acquired heart disease. On Friday afternoon, a panel of leading radiologists will extend this discussion to radiologic diagnosis and compare the value of cinefluorographic and conventional techniques in the functional and anatomic diagnosis of cardiovascular diseases. Proffered cinefluorograms demonstrating new findings, techniques and apparatus will be shown on Friday evening.

Saturday morning will be devoted to cinefluorographic equipment; motion-picture technique, including editing, print control, and special effects; cinefluorographic installations; and miscellaneous equipment such as recording devices and injection apparatus. Saturday afternoon is reserved for the presentation of submitted papers dealing with previously unpublished information on radiologic findings of the cardiovascular system, the gastrointestinal tract, urinary tract, etc., as demonstrated by cinefluorography. Presentation of these papers will be limited to 20 minutes plus an additional 10 minutes for discussion.

Anyone who wishes to show a cinefluorogram on Friday evening must send a short description of the material covered, footage, number, make and size of the image tube used. Also, those individuals desiring to present a paper on Saturday afternoon must submit a 500-word abstract. The deadline for the descriptions of cinefluorograms and abstracts of papers is September 1, 1959. In order to provide opportunity for review by the Symposium Committee, a copy of the paper to be presented must be forwarded to Dr. George H. Ramsey, Professor and Chairman, Dept. of Radiology, Strong Memorial Hospital, Rochester 20, N. Y., by October 1, 1959. It should be understood that the papers and accompanying discussions will become the property of the Symposium Committee who will arrange for their editing and publication. The preliminary program of the Symposium will be mailed in the early Fall.

## COURSE IN RADIOACTIVE ISOTOPES FOR TECHNICIANS

A course in radioactive isotopes for technicians will be held at Queens Hospital Center, Department of Hospitals, The City of New York. The course will consist of 8 sessions of 4 hours each, to be held every Wednesday afternoon, commencing September 30, 1959, and ending November 25, 1959 (1 to 5 P.M.). The enrollment is limited to 10 members, and the tuition is $100.00.

Requests for applications should be sent to: Dr. Philip Kahan, Supervising Medical Superintendent, Queens Hospital Center, 82–68 164th Street, Jamaica 32, New York.

# BOOK REVIEWS

*Books sent for review are acknowledged under: Books Received. This must be regarded as a sufficient return for the courtesy of the sender. Selections will be made for review in the interest of our readers as space permits.*

THE CHEMICAL PREVENTION OF CARDIAC NECROSES. By Hans Selye, M.D., Ph.D., D.Sc., Professor and Director of the Institute of Experimental Medicine and Surgery, University of Montreal, Canada. Cloth. Pp. 235, with illustrations. The Ronald Press Company, New York, N. Y., 1958.

In this very readable book the author describes his investigations to disclose the common pathway by which the sequence of necrosis, inflammation and fibrosis is produced in the heart muscle.

From his experimental work with animals and the observations of other workers, he develops the theory that, after conditioning with certain corticoids, pathogens and other factors, myocardial lesions may be produced quite regularly, the lesions depending largely upon the anion. To quote directly from the book: "When NaCl is used following conditioning with corticoids the change is termed ESCH (electrolyte-steroid-cardiopathy with hyalinization), characterized mainly by hyalinization and periarteritis nodosa of the cardiac vessels. Other Na-salts, the phosphates, sulfates and perchlorate, induce a necrotizing cardiopathy, the ESCN (electrolyte-steroid-cardiopathy with necrosis). This latter change, unlike the former, is readily prevented by prophylactic administration of KCl or $MgCl_2$."

The author has coordinated the isolated clinical and experimental observations in the world literature with personal knowledge derived from his animal experimentation. The result is a most interesting and stimulating formulation of a concept of the interplay between electrolytes, steroids and stress, a concept that deserves the serious attention of those concerned with the prevention of cardiac necroses.

WILLIAM S. REVENO, M.D.

AVIATION MEDICINE—SELECTED REVIEWS. Edited by Clayton S. White, W. Randolph Lovelace II, and Frederic G. Hirsch, The Lovelace Foundation for Medical Education and Research, Albuquerque, New Mexico. Cloth. Price $12.00. Pp. 305, with numerous illustrations. Pergamon Press, Inc., 122 East 55th Street, New York 22, New York, 1958.

Some readers may find the title somewhat misleading since this book is primarily a scholarly contribution assessing instrumentation and related methodology in Physiology.

The subjects treated are: Spirometric Methods, by Ulrich C. Luft; High-Speed Motion Picture Photography as an Aid in Bio-Medical Investigation, by Merrill A. Palmer; The Measurement of Atmospheric Ozone, by I. Gerald Bowen; Methods and Apparatus for the Study of Stress Reactions and Metabolic Changes, by Bernard B. Longwell; Dosimetry of Ionizing Radiations, by John L. Howarth; Some Technical Developments Applicable to Problems in Pathology, by T. L. Chiffelle; Temperature Measuring Techniques for Aviation Physiological Research, by Frederic G. Hirsch; The Analysis of Respiratory Gases, by Clayton S. White; Aerosols: Physical Properties, Instrumentation and Techniques, by Arnold E. Reif; Pressure Transducers—Transformer Type, by James Clark; Spectrometric Methods: Gas Analysis by Quantitative Emission Spectroscopy, by Clayton S. White and W. Randolph Lovelace II; Comparison of Rahn-Otis Technique with other Methods of Intermittent and Continuous Sampling of Expired Air, by Nils P. V. Lundgren; and Clayton S. White presents a summary of the main ideas and objectives of each review.

The book is handsomely bound; the photographs are impressive, being of the high contrast kind; the diagrams are superb; the references are extensive and include the title of each article; and the 28 page index is well done. There is nothing superficial about this volume.

WALTER H. SEEGERS, PH.D.

THE UPPER ATMOSPHERE. By H. S. W. Massey, Ph.D., D.Sc., F.R.S., Quain Professor of Physics, University College, London, and R. L. F. Boyd, Ph.D., Lecturer in Physics, University College, London. Cloth. Price, $17.50. Pp. 333, with 4 full page color illustrations and 21 in black and white. Philosophical Library, Inc., 15 East 40th Street, New York 16, N. Y., 1959.

This is an unusual achievement in science exposition. There is nothing in the book requiring

advanced knowledge but the volume certainly contains an authoritative coverage of the subject. This account makes it possible for self-education in a direction where there is so much popular discussion today. Here are hard facts without the glamour, presented in a style that is charming in its own right.

Following a discussion of relevant physics, there are eleven more chapters discussing the atmosphere; research by balloons and rockets; probing with sound waves; probing with radio waves; the ozonosphere and the ionosphere; lights in the night sky; aerial tides and magnetic effects; solar, magnetic, and ionospheric disturbances; meteors; cosmic rays; and artificial satellites. A concluding chapter covers future possibilities.

A desire to extend the general knowledge in this field is the spirit that permeates this book rather than its military aims. The many charts show free hand lettering and give the book a certain softness that fits the whole format so well. Although treatment of the subject is comprehensive, there are no detailed references and only a short bibliography.

Walter H. Seegers, Ph.D.

## BOOKS RECEIVED

Cliniques Radiologiques. Première Série. L'agrandissement Radiographique Direct Dans L'examen de la Base du Crane et des Micro-Fractures du Rocher. By Professeur J. Roussel, with collaboration of P. Schoumacher, M. Pernot, and R. Poiré. Paper. Price, 3,200 fr. Pp. 102, with 59 illustrations. Masson et C^ie^, Éditeurs, 120 Boulevard Saint-Germain, Paris, France, 1959.

Angiocardiopneumographie Élargie. Méthode D'opacification Vasculaire Générale Par Voie Veineuse. By P. Viallet, Électroradiologiste des Hôpitaux (Alger), L. Sendra, Chef de Clinique à la Faculté de Médecine d'Alger, L. Chevrot, Ancien Chef de Clinique à la Faculté de Médecine d'Alger, P. Aubry, Chef de Clinique à la Faculté de Médecine d'Alger and P. Combe, Professeur agrégé à la Faculté de Médecine d'Alger. Paper. Price, 4,400 fr. Pp. 110, with 46 illustrations. Masson et C^ie^, Éditeurs, 120 Boulevard Saint-Germain, Paris, France, 1959.

Atlas of Roentegenographic Measurement. By Lee B. Lusted, M.D., Associate Professor of Radiology, University of Rochester School of Medicine and Dentistry, and Theodore E. Keats, M.D., Professor of Radiology, University of Missouri School of Medicine. Cloth. Price, $9.00. Pp. 176, with 119 illustrations. The Year Book Publishers, Inc., 200 East Illinois Street, Chicago, Illinois, 1959.

An Atlas of Normal Radiographic Anatomy. Second edition. By Isadore Meschan, M.A., M.D., Professor and Director, Department of Radiology, Bowman Gray School of Medicine of Wake Forest College, Winston-Salem, North Carolina; Consultant, Walter Reed Army Medical Center, Washington, D. C.; Formerly Professor and Head of the Department of Radiology, University of Arkansas School of Medicine, with the assistance of R. M. F. Farrer-Meschan, M.D., B.S., M.D. (Melbourne, Australia), Research Associate, Department of Radiology, Bowman Gray School of Medicine of Wake Forest College, Winston-Salem, North Carolina. Cloth. Price, $16.00. Pp. 759, with numerous illustrations. W. B. Saunders Company, 218 W. Washington Sq., Philadelphia, Pennsylvania, 1959.

Low Intensity Radium Therapy. By Charles L. Martin, M.D., Clinical Professor of Radiology, Southwestern Medical School, University of Texas, and James A. Martin, M.D., Associate Professor of Radiology, Southwestern Medical School, University of Texas. Cloth. Price, $12.50. Pp. 277, with 138 illustrations. Little, Brown & Company, 34 Beacon St., Boston, Massachusetts, 1959.

Principles of Radiation Dosimetry. By G. N. Whyte, National Research Council, Ottawa, Canada, and Queen's University, Kingston, Ontario. Cloth. Price, $7.00. Pp. 124, with numerous illustrations. John Wiley & Sons, Inc., 440 Fourth Ave., New York, New York, 1959.

Radiation Therapy. By Walter T. Murphy, M.D., Director of Therapeutic Radiology, Roswell Park Memorial Institute, Buffalo, New York; Consultant in Radiation Therapy, U. S. Veterans Hospital, Buffalo, New York; Associate Clinical Professor of Radiology, University of Buffalo Medical School. Cloth. Price, $25.00. Pp. 1,041, with numerous illustrations. W. B. Saunders Company, 218 W. Washington Sq., Philadelphia, Pennsylvania, 1959.

Evaluation of the Pelvis in Obstetrics. Second edition. By Charles M. Steer, M.D., Med.Sc.D., F.A.C.S., F.A.C.O.G., Associate Professor of Clinical Obstetrics and Gynecology, College of Physicians and Surgeons, Columbia University, and the Sloane Hospital for Women. Cloth. Price, $4.00. Pp. 131, with 57 illustrations. W. B. Saunders Company, 218 W. Washington Sq., Philadelphia, Pennsylvania, 1959.

Cerebral Angiography in the Management of Head Trauma. American Lecture Series. By Charles A. Carton, M.D., Chief, Division of Neurosurgery, Montefiore Hospital; Associate Clinical Professor of Surgery (Neurosurgery), Albert Einstein College of Medicine, New York. Cloth. Price, $7.00. Pp. 157, with 44 illustrations. Charles C Thomas, Publisher, 301–327 East Lawrence Ave., Springfield, Illinois, 1959.

Vol. 82, No. 3

# ABSTRACTS OF RADIOLOGICAL LITERATURE

*Department Editor:* T. Leucutia, M.D., Harper Hospital, Detroit 1, Michigan

## INDEX TO ABSTRACTS

### ROENTGEN DIAGNOSIS

#### Genitourinary System


Mathews, P. W., Jr.: Experience with renografin 60%, a urographic medium ..... 568

Orr, L. M., Campbell, J. L., and Thomley, M. W.: Intravenous urography with renografin 60%; a report of more than 600 cases. 568


#### Nervous System


MacCarty, C. S., Leavens, M. E., Love, J. G., and Kernohan, J. W.: Dermoid and epidermoid tumors in the central nervous system of adults ..... 568

Reinhardt, K.: Observations in accidentally produced thoracic abrodil-myelography ..... 569

Ganado, W., and Craig, A. J.: Brucellosis myelopathy ..... 569

Nordlander, S., Salén, E. F., and Unander-Scharin, L.: Discography in low back pain and sciatica; analysis of 73 operated cases ... 569


#### Skeletal System


Clausen, H. J.: An individual with twenty-five presacral vertebrae and thirteen pairs of ribs ..... 570

Crow, N. E., and Brogdon, B. G.: The "normal" lumbosacral spine ..... 570

Gillman, E. L.: Congenital absence of the odontoid process of the axis; report of a case. 570

Haveson, S. B.: Congenital flatfoot due to talonavicular dislocation (vertical talus) ..... 570

Vickers, C. W., Pugh, D. C., and Ivins, J. C.: Osteoid osteoma; a fifteen-year follow-up of an untreated patient ..... 571

Jacobson, S. A.: Early juxtacortical osteosarcoma; (parosteal osteoma) ..... 571

James, C. C. M., and Lassman, L. P.: Diastematomyelia ..... 572

Maroteaux, P., and Lamy, M.: Pseudo-achondroplastic forms of spondylo-epiphyseal dysplasias ..... 572

Glenchur, H., Zinneman, H. H., and Hall, W. H.: A review of fifty-one cases of multiple myeloma ..... 572

## ROENTGEN DIAGNOSIS

### GENITOURINARY SYSTEM

MATHEWS, P. W., JR. Experience with renografin 60%, a urographic medium. *South. M. J.*, Feb., 1959, *52*, 170–173. (From: The Department of Radiology, North Carolina Baptist Hospital, Inc., Winston-Salem, N. C.)

The author has conducted a very careful survey of 432 patients who were examined with renografin 60 per cent. He found that a satisfactory diagnosis could be made in 68.8 per cent of these patients and a limited diagnosis in 23.4 per cent for a total of 92.2 per cent. On the basis of these findings, he concludes that this substance is a very effective contrast medium for delineating the upper urinary tract.

A careful check of allergy histories was kept. Forty-four of the patients examined had a history of allergy. It is interesting to note that the incidence of side reactions was higher in those patients with a history of allergy than in those with no history of allergy. The author states that intradermal or intravenous tests were done on all patients prior to the examination. He points out that there was no apparent correlation between the occurrence of a positive reaction to the intravenous test and the occurrence of a side reaction during the examination. This is in apparent agreement with other reports which indicate that most of the preliminary testing done is not very helpful in deciding who will react to the intravenous contrast medium used.—*Richard E. Kinzer, M.D.*

ORR, LOUIS M., CAMPBELL, JAMES L., and THOMLEY, MILES W. Intravenous urography with renografin 60%; a report of more than 600 cases. *J.A.M.A.*, March 14, 1959, *169*, 1156–1158. (Address: L. M. Orr, 1300 Kuhl Ave., Orlando, Fla.)

The authors used 60 per cent renografin in 654 consecutive cases over a period of one and one-half years; the results are the purpose of this report. Sixteen of the patients had a history of hay fever, asthma or severe reaction to previously administered contrast agents. Because results on a previous series with renografin by intravenous injection in 1 cc. amounts revealed no reaction, the authors did not do intravenous testing in this group of patients.

Each patient was prepared with 12 fluid ounces of magnesium citrate solution thirty-six hours before the examination, with light eating the day before and nothing by mouth after midnight except coffee and toast for breakfast on the day of examination. Twenty-five cc. of 60 per cent renografin was given intravenously over a period of three minutes with smaller amounts being given only in 3 cases of younger individuals. Roentgenograms were taken at five, ten, and fifteen minutes, the last roentgenogram with the patient in an upright position.

There were no severe reactions in any patient, but 40 of the patients reported a total of 58 different mild reactions, the majority of which were nausea, vomiting, flushing of the skin with occasional choking sensation, tingling of the extremities and itching, urticaria or sneezing.

In rating the examinations in terms of excellent, good, or fair, good or excellent results were obtained in 97 per cent of the cases. The authors state that, in their hands, renografin 60 per cent has been a medium which has produced excellent roentgenograms and remarkably few and mild reactions.—*Stephen Bruny, M.D.*

### NERVOUS SYSTEM

MACCARTY, COLLIN S., LEAVENS, MILAM E., LOVE, J. GRAFTON, and KERNOHAN, JAMES W. Dermoid and epidermoid tumors in the central nervous system of adults. *Surg., Gynec. & Obst.*, Feb., 1959, *108*, 191–198. (From: The Sections of Neurologic Surgery and Pathologic Anatomy, Mayo Clinic and Mayo Foundation, Rochester, Minn.)

The authors consider epidermoid tumors as congenital in origin and as comprised of the outer layer of the epidermis, squamous epithelium, and desquamated epithelium. Dermoid tumors are teratomas composed primarily of skin and its derivatives. The tumors discussed in this paper occurred for the most part in adults.

A total of 44 epidermoid tumors of the central nervous system were encountered at the Mayo Clinic before December 1955. Twenty-four occurred intracranially; 14 were in the skull; 3 were in the orbital area; and 3 were intraspinal in location. The tumors in the intraorbital area were removed by frontal craniotomy and the results were good with no recurrences.

Of the 14 patients with tumors involving the skull, palpable tumors were evident in 10 cases. Roentgenographic studies of the skull in these cases usually showed involvement of both tables by a smooth, scalloped, and sclerotic margin around an area of radiolucency. In some cases, both tables of the skull were destroyed. These cases were all treated surgically with relatively good results.

The intracranial epidermoid tumors were most commonly found in the cerebellopontine angle, sellar or temporal lobe area.

Three adult patients were seen with intraspinal epidermoid tumors. These 3 patients have survived. All 3 had either positive myelograms, or there was evidence of erosion of the spinal column or a vertebra.

Thirty-two dermoid tumors were recorded in this series. Six were located intracranially in the supratentorial area and 3 were located intracranially in the infratentorial area. There were 7 intradiploic dermoid

tumors found in children and 4 in adults. One patient was seen with an intraorbital tumor. Five patients were found with spinal dermoid tumors.

Dermoid tumors presented calcification much more frequently than the epidermoid tumors.

Congenital anomalies were seen in 50 per cent of the dermoid tumors while only 3 of the epidermoid tumors were associated with congenital anomalies. A large number of patients with epidermoid as well as with dermoid tumors had mental symptoms.—*Richard E. Kinzer, M.D.*

Reinhardt, K. Zufällige Beobachtungen von unteren thorakalen Abrodilmyelographien. (Observations in accidentally produced thoracic abrodil-myelography.) *Radiol. clin.*, Jan., 1959, *28*, 49–60. (From: Strahleninstitut der Universitätskliniken im Landeskrankenhaus, Homburg Saar, Germany.)

Prior to the particular experience under discussion, the author avoided the use of water soluble iodine preparations in thoracic myelography because of the annoying side reactions or serious complications. He was therefore surprised when abrodil in the thoracic region was so well tolerated. The accidental flow into the thoracic portion of the subarachnoid space occurred in a patient who had a peridural lymphogranuloma with compression of the cauda equina.

Abrodil is preferred to the iodized oils because it shows subtle pathologic changes more clearly.

Satisfactory anesthesia was secured by the injection of 4 cc. novocaine into the lumbar region. Adequate myelograms were obtained with 10 cc. abrodil. The same substance could be used for peri- and subdurography.—*J. Zausner, M.D.*

Ganado, Walter, and Craig, Alfred John. Brucellosis myelopathy. *J. Bone & Joint Surg.*, Dec., 1958, *40-A*, 1380–1388. (From: Royal University of Malta and St. Luke's Hospital, Valletta, Malta, G. C.)

Brucella may cause meningomyelitis, or local arachnoiditis and resultant paraplegia. However, it may also produce paraplegia as a complication of spondylitis secondary to cord compression.

Brucellosis spondylitis simulates tuberculous spondylitis. The former is often characterized by an insidious onset with mild symptoms and an indolent course. From 1947 to 1954, 5,600 cases of brucellosis were reported in Malta, and 130 had brucellosis spondylitis. Ten of these had signs and symptoms of compression myelopathy. Brucella organisms are rarely found in the blood, since myelopathy develops late in the course of the disease.

There is a granulomatous reaction to Brucella with tubercles and caseation; very small nodules are formed. The lesions are more extensive than they appear on the roentgenograms and skeletal structures apparently negative on the roentgenograms may be involved.

Initially, the lesions of the spinal column are in the subchondral bone and, when the cartilage is destroyed, the intervening disk is devitalized but shows no inflammatory reaction. The vertebral bodies tend to collapse and the disk narrows. Osteophytes form around the margins of the vertebral bodies and tend to bridge the disks. These osteophytes form very quickly, usually within two to three months. Cold abscesses may form. Compression of the cord occurs as in tuberculosis.

Six cases are reported; 5 exhibited the typical changes in the spine, but one had no abnormalities noted on the roentgenograms. Surgery in this case showed an epidural abscess, apparently from a small spinal focus as suppuration has never been reported from primary meningeal Brucella infection.

The prognosis is good when rest and antibiotics are prescribed early.—*Martha Mottram, M.D.*

Nordlander, Sverker, Salén, Ernst F., and Unander-Scharin, Lars. Discography in low back pain and sciatica; analysis of 73 operated cases. *Acta. orthop. Scandinav.*, 1958, *28*, 90–102. (From: Orthopaedic Department and the Department of Diagnostic Roentgenology, Karolinska Institutet, and the Roentgen Department, Norrbackainstitutet, Stockholm, Sweden.)

Between 1948 and 1958 more than 500 diskographic examinations have been performed at the Roentgen Diagnostic Department of Karolinska Sjukhuset. Since the myelographic examination cannot disclose all types of disk prolapse, either because of the size of herniation or the location, the authors have analyzed 73 operative cases, most of which had both myelographic and diskographic examination. The reason for this study was the frequency of prolapsed disk found surgically despite a negative myelogram.

Evaluation of the findings of diskography is based on the roentgen analysis of the disk space and on reproduction of the pain by the puncture of the posterior longitudinal ligament and distention of the disk space with the contrast medium. The authors believe that the reproduction of the pain is not due to a chemical irritant but rather to a mechanical one and they quote an article by Hirsch as proving this. The authors state that, despite the reproduction of the typical pain on injection, there was no uniformity in response in their series of patients and that evaluation of the response was an extremely difficult task.

The pathologic roentgenologic findings are as follows: (a) The normal disk shows either a well delineated space usually lying somewhat dorsally in the disk space area, or two areas of opacification, in layers, and usually connected by a narrow central channel; (b) severe disk degeneration is manifested

by flattening and elongation of the disk, with irregular outline; (c) dorsolateral rupture without leakage is associated with an enlarged disk space and a more or less sharply delimited streak of contrast medium posteriorly or posterolaterally. The ruptured disk would be demarcated, if leakage is present, by the accumulation of contrast medium under the posterior longitudinal ligament; (d) epidural leakage usually indicates degeneration of the disk space associated with a well defined posterior perforation of the posterior longitudinal ligament and communication with the epidural space, as demonstrated by the streaks of contrast medium around the dural sac; (e) the perineural leakage of contrast medium around the nerve roots in the intervertebral foramina likewise indicates rupture and degeneration.

The authors conclude that there is no question of demonstrating the superiority of diskography over myelography, but rather that the two methods may complement each other. In cases of low back pain or sciatica when the myelogram is negative, diskography may be indicated, if the signs and symptoms so warrant. They also state that the cases of chronic low back pain with negative myelograms but with an indication for surgery, and cases for investigation, such as insurance cases, with obscure pain do warrant evaluation by diskography.—*Stephen Bruny, M.D.*

## SKELETAL SYSTEM

CLAUSEN, H. J. An individual with twenty-five presacral vertebrae and thirteen pairs of ribs. *Anat. Rec.*, Aug., 1958, *131*, 737–743. (From: Department of Anatomy, University of Arkansas Medical Center, Little Rock, Ark.)

Several skeletal variations were found in a one-hundred-four year old male Negro cadaver during the course of routine laboratory dissections in gross anatomy. The cause of death was indicated as bronchopneumonia. The vertebral column was peculiar in that there were 25 presacral vertebrae. The twentieth vertebra carried the thirteenth pair of ribs. One of these ribs articulated with the body, pedicle and the transverse process of the vertebra. Therefore, this pair of ribs was considered to be a true thoracic pair of ribs which articulated with a characteristic thoracic vertebra.

A subcostal and 12 intercostal pairs of nerves were also observed.

The skeletal variations in the thoracic region of this specimen might well be accounted for by an irregularity in costal segmentation.—*Thomas R. Marshall, M.D.*

CROW, NEIL E., and BROGDON, BYRON G. The "normal" lumbosacral spine. *Radiology*, Jan., 1959, *72*, 97. (Address: N. E. Crow, USAF Hospital, Lackland AFB, Texas.)

The authors present a statistical review of the roentgenograms of the lumbosacral spine of 936 healthy young males, utilizing the double reading technique. The men were asymptomatic Air Force Academy or Air Cadet applicants between the ages of seventeen and twenty-seven. The results are presented in tabular form.

This study disclosed the presence of spina bifida occulta in 35.7 per cent of the series, previous Scheuermann's disease in 20.7 per cent, and transitional vertebrae in 11.5 per cent. Other conditions which were less frequently observed include scoliosis, rudimentary ribs, spondylolysis, spondylolisthesis, limbus vertebrae and hemangioma. It is of interest that only 40.2 per cent presented none of the above variations.—*Edward B. Best, M.D.*

GILLMAN, E. L. (Long Beach, Calif.) Congenital absence of the odontoid process of the axis; report of a case. *J. Bone & Joint Surg.*, March, 1959, *41-A*, 345–348.

Prior to this case report, 21 cases of congenital malformation of the dens epistrophei have been described, the first report being in 1933. These anomalies have been classified into three types: total absence of the dens; partial absence (hypoplasia); and failure of fusion of the tip or separate ossicle (os odontoideum).

The author reports another case of congenital aplastic odontoid process of the axis. A blow on the head was followed by symptoms of distress low in the neck; these were mild and not disabling. At no time while under clinical observation did the patient have symptoms referable to the defective, unstable atlanto-axial joint. It is pointed out by the author that instability of the atlanto-axial joint, due to anomaly of development, in itself presents a less grave hazard to health or life than one would expect.

Treatment is discussed. In the 21 cases reported there were 3 deaths. One appeared to be the result of surgical intervention. In another, pronounced platybasia with malformed occipital lobes of the brain and a Klippel-Feil deformity were found at autopsy. Death was due to tonsillar herniation into the foramen magnum. In the third death, the patient had complained of headache and vertigo for one month, with posterior dislocation of the atlas. An autopsy was not performed. Fusion was recommended to 10 patients, but performed in only 3. The majority of the reported patients refused the risk of fusion which, to them, seemed to outweigh the clinical severity of the symptoms. In general, treatment was limited to the usual precautions against serious trauma to the head, such as might occur in diving, tumbling or wrestling. Other than prophylactic indications are necessary before surgical fusion of the joint is performed. Reproductions of roentgenograms and laminagrams are included.—*Douglas S. Kellogg, M.D.*

HAVESON, SAMUEL B. Congenital flatfoot due to talonavicular dislocation (vertical talus). *Radiology*, Jan., 1959, *72*, 19–25. (Address:

University of California Medical Center, San Francisco, Calif.)

Congenital talonavicular dislocation is an infrequent anomaly causing flatfoot. Few cases have been reported and little information concerning this deformity can be found in the orthopedic text books or in the English radiologic literature.

The etiologic factors resulting in the anatomic derangement are not known with certainty. It is assumed that the distal portions of the talus and calcaneus rotate toward the plantar surface. This results in medial rotation of the talus and eversion of the calcaneus at the subtalar joint. The vertical position of the talus allows only the posterior part of its superior articular surface to articulate with the tibiofibular mortice. A dislocation of the talonavicular joint results and there is relative dorsiflexion of the forefoot at the transverse tarsal joints and obliteration or reversal of the longitudinal arch. Shortening of the Achilles tendon is a frequent finding. Whether this shortening is congenital or represents an adaptive process is not known.

At birth, the deformity of the foot may be sufficiently marked to attract the attention of the examining physician or parent. The forefoot is turned outward and there is a prominent bulge on the plantar surface at the midtarsal region. The diagnosis in the older child and adult is easily made by roentgen examination. A lateral projection of the foot reveals the characteristic vertical talus, the articulation of the navicular with the superior surface of the neck of the talus and rotation of the distal calcaneus toward the plantar surface. Medial displacement of the distal talus and abduction of the forefoot are demonstrated in the dorsoplantar projection.

Four cases of this anomaly are presented.—*Arno W. Sommer, M.D.*

Vickers, Charles W., Pugh, David C., and Ivins, John C. Osteoid osteoma; a fifteen-year follow-up of an untreated patient. *J. Bone & Joint Surg.*, March, 1959, *41-A*, 357–358. (From: The Sections of Roentgenology and Orthopaedic Surgery, Mayo Clinic and Mayo Foundation, Rochester, Minn.)

This is a report of a fifteen year follow-up of an untreated patient. From the time of its original description as a separate disease entity, osteoid osteoma has been considered to be a surgically curable disease, and most of the reported cases have been treated by excision of the lesion. Six cases have been described in which untreated osteoid osteomata have been observed from the painful phase until the patient was relatively symptom-free.

In the current case, the patient was first seen in 1943 and his chief complaint was a deep boring pain of diffuse distribution in the right thigh and low in the back, of five years' duration. A roentgenogram of the upper portion of the right femur revealed a fusiform thickening of the cortex in the subtrochanteric region in the center of which was a nidus 1 cm. in diameter. The lesion was considered to be either an osteoid osteoma or osteomyelitis, and surgical treatment was recommended. The patient agreed to return for the operation at his earliest convenience. His next visit to the clinic was in 1958. The history indicated that the pain had persisted for the entire fifteen year period but had been diminishing progressively, and at the time of his second visit he was almost symptom-free. A roentgenogram of the upper part of the right femur showed little change in the appearance of the lesion. In view of the fact that the patient was almost symptom-free, no definitive treatment was recommended. After reviewing the history and roentgenograms, it was agreed that the lesion was an osteoid osteoma.

Since the lesion regressed spontaneously in all of the reported cases, and almost certainly does so in all untreated cases, one might question the theory that the lesion is by nature a true neoplasm. The self-limiting characteristics of the condition would tend to place it in the category of an inflammatory lesion. The microscopic picture is not, however, one of infection. In view of our present knowledge, it is not possible to place the disease in any specific category.—*Douglas S. Kellogg, M.D.*

Jacobson, Sheldon A. Early juxtacortical osteosarcoma; (parosteal osteoma). *J. Bone & Joint Surg.*, Dec., 1958, *40-A*, 1310–1328. (From: Bone Tumor Registry, Department of Pathology, University of Oregon Medical School, Portland, and the United States Veterans Administration Hospital, Vancouver, Wash.)

The author reports 8 cases of parosteal osteoma or juxtacortical osteosarcoma. Four of these were phalangeal tumors, 3 femoral and 1 vertebral. In the previous reported series of Geschickter and Copeland, and Dwinnell, Dahlin and Ghormley, some of the cases of juxtacortical osteosarcoma were originally classified as osteochondroma or exostosis.

The author suggests that there are some lesions that appear to be merely osteochondroma superficially, but can be differentiated from them by their histologic appearance, a tendency to recurrence and occasional malignant degeneration into juxtacortical osteosarcoma. Possibly they should be considered to be premalignant lesions.

Histologically, there is marked cellularity of the base of the lesion as well as of the capsule with cellular pleomorphism and hyperchromatism. There is cellular fibrous connective tissue within the stalk of the osteochondroma rather than fatty tissue. Areas of unspecialized connective tissue, cartilage and osteoid, with no recognizable polarity, in the cap are believed to be pathognomonic of this lesion. They may even show no cartilage cells. Growth may proceed both away from and into bone.

It is not definite that simple resection is followed by malignant recurrence. It is suggested that wide

excision be done first and, if there is recurrence, an aggressive surgical procedure be undertaken. If it is confirmed that these tumors develop into sarcoma, a recurrence should constitute an indication for amputation whenever this is feasible.—*Martha Mottram, M.D.*

JAMES, C. C. MICHAEL, and LASSMAN, L. P. Diastematomyelia. *Arch. Dis. Childhood*, Dec., 1958, *33*, 536–539. (From: Departments of Orthopaedic Surgery and Neurological Surgery, Newcastle upon Tyne General Hospital, Newcastle upon Tyne, England.)

A case of diastematomyelia is reported which was treated surgically with excellent results. A girl of ten years had poor leg circulation, a slightly small right foot and pain in the right foot. Roentgenograms showed spina bifida from L3 to S1, a midline bone spur L2 to L3 and a division of the oil column on myelography. Nine months after removal of the bony and fibrous septum, no abnormalities remained aside from a slight disturbance of circulation in the feet.

Diastematomyelia is usually recognized in children or young adults. Progressive weakness in the legs or deterioration of the gait suggests the diagnosis.

This condition usually is present in the lower thoracic or lumbar areas but has been reported as high as T2. A septum of bone, fibrous tissue, or fibrocartilage divides the spinal cord longitudinally. This septum fixes the cord and divides the neural canal anteroposteriorly in the midline. It is often found in association with spina bifida, myelocele, or dermoid. Since the septum fixes the cord, the subsequent clinical effects become more marked with growth as the cord ascends. This ascent appears to be most rapid around puberty. If the tension on the cord nerves or blood vessels is considerable, there will be progressive neurologic changes, unequal size of the legs, incontinence, paresis, or trophic changes.

A bony septum is often demonstrated on roentgenography, but in young children it may not be sufficiently calcified to be identified. If suspected clinically, myelograms or repeat examinations should be done. Spina bifida or increased interpedicular distance, without pedicular erosion, are suggestive of diastematomyelia.

The authors stress that early diagnosis and surgery are important when the cord is fixed by a septum, as there is increasing tension resulting in progressive symptoms with growth.—*Martha Mottram, M.D.*

MAROTEAUX, PIERRE, and LAMY, MAURICE. Les formes pseudo-achondroplasiques des dysplasies spondylo-épiphysaires. (Pseudo-achondroplastic forms of spondylo-epiphyseal dysplasias.) *Presse méd.*, Feb. 25, 1959, *67*, 383–386. (From: Clinique de Génétique médicale, Hôpital des Enfants-Malades, Paris, France.)

Three cases of spondylo-epiphyseal dysplasia (probably genotypic recessive) are presented and the world literature is summarily reviewed. These simulate achondroplasia; however, a thorough study reveals four important differences. Whereas achondroplasia is discovered in infancy, these patients show signs of abnormality at and after the age of two. The shortening of the extremities, in particular of the hands, is more pronounced. No clinical and roentgenographic abnormality of the face or of the skull can be noted. Also, marked epiphyseal and metaphyseal changes (late appearance, irregularity and fragmentations of epiphyses) as well as vertebral deformities (narrow, biconvex, elongated and at times cuneiform vertebral bodies) are demonstrated roentgenographically; hence, resemblance of these cases to Morquio's disease is noted—*Jirair N. Sarian, M.D.*

GLENCHUR, HARRY, ZINNEMAN, HORACE H., and HALL, WENDELL H. A review of fifty-one cases of multiple myeloma. *A.M.A. Arch. Int. Med.*, Feb., 1959, *103*, 173–183. (From: Veterans Administration Hospital and the Department of Medicine, University of Minnesota, Minneapolis, Minn.)

Experiences with 51 consecutive cases of multiple myeloma are reported with emphasis on the incidence of infection, particularly bacterial pneumonia. This study was prompted by reported observations that recurrent bacterial infections occur in patients with agammaglobulinemia; that patients with multiple myeloma are susceptible to recurrent bacterial infection; and that certain patients with multiple myeloma and large amounts of abnormal serum globulins have little or no gamma globulin or serum antibody.

Results of the review are in agreement with previous reports. There were 18 cases with myelomatous kidneys, 10 cases presented with paraplegia, and 2 with rheumatoid arthritis had amyloidosis.

Fifty-five per cent of 51 patients experienced one or more bouts of pneumonia (exclusive of terminal pneumonia), averaging 2.7 bouts per patient, and 17 had terminal pneumonia. Fifty-seven per cent had other infections, predominantly of the urinary tract.

Only 1 of the 55 patients had a normal serum electrophoresis pattern.—*Lois Cowan Collins, M.D.*

JAMES T. CASE

Janeway Lecturer, 1959